PRESENT STATE OF CHILD NEUROLOGY

*Introductory Lecture at the N.S.S. Oxford Study Group on Child Neurology and Cerebral Palsy**

CYRILLE KOUPERNIK.

From the Salpêtrière Hospital and the International Children's Centre, Paris.

Child Neurology has changed in the last 25 years. When one reads a textbook of the early thirties one cannot but notice that the techniques used are those of adult neurology, and that the main factor inherent to the child—i.e., development—is not taken into account. The second difference lies in the progress accomplished in the purely technical field; means of investigation that were used 25 years ago were, of course, considerably poorer than ours. A third point I want to stress is the general attitude towards the crippled child, made up of rejection and therapeutic abstention. There was a time when almost all the cerebral palsies and encephalopathies were systematically explained in terms of hereditary lues and degeneration; you can easily discover behind these words, these pseudo-scientific concepts, the deeply-rooted belief that the spastic is a different person and that he is such because his parents or grandparents have sinned. The battle is lost from the very beginning; the word "rehabilitation" is not even used.

When we come to summarising the progress accomplished in these 25 years, we must admit that though this particular field of Medicine has benefited from the general evolution of our medical knowledge, its progresses have probably been less striking than, for instance, those in infectious diseases. Furthermore, the revolution brought about by antibiotics has even complicated our own task—I mean that before they existed many young spastics could not survive and that now we probably have to deal with even more bad cases.

Normal to Abnormal

In dealing with cerebral palsy the pædiatrician has to solve two problems: (1) to recognise the condition early, and (2) to try to understand the underlying process. I would like to discuss the second point first. It has been a well-admitted attitude in Medicine to explain normal functioning by pathology. But with the development of biological disciplines the trend is more and more to begin with physiology—i.e., with normal function—and to try to understand disease in terms of health. This means that if we want to understand what goes wrong with the development of the cerebral palsied child we have first to know how the normal infant develops. This is the first and a very important reason why we infant neurologists must first study the development of the normal infant. Let us come back now to the first point I stressed, that is, the early recognition of the abnormal condition. André Thomas told me only two days ago that when at the age of 76 he decided to study the infant he found the techniques of adult neurology absolutely inadequate and had to devise new ones. You know, for instance, that very often such a severe condition as kernicterus is not unmasked during the first five or six months, though it is obviously there since birth or even before. So, we have to face the necessity of this particular type of knowledge, that of the normal infants at a given age.

*Held at St. Edmund Hall, Oxford, from September 21 to 27, 1958. A report by Dr. Mary Capes, a group photograph, and three papers on Body Image, read at the meeting, appeared in Bulletin No. 4.—Ed. C.P. Bulletin.

I obviously cannot make a full historical survey here and I will refer only to the greater pioneers in this field, my teacher Arnold Gesell, Myrtle MacGraw and André Thomas, this disciple of Babinski, who, as I already told you, started in the late seventies, at an age when other people have retired, to study the neonate, because he wanted to understand how the anencephalic behaves. He is still, at over ninety, at work, mainly with our friend Minkowski's team and especially with Madame Saint-Anne Dargassies, whose film Minkowski will show you this week.

Gesell had to endure the reproach of not paying sufficient attention to the child's milieu. I am not going to resume this eternal argument of Nature *v.* Nurture; all I want to say is that there is good evidence proving that maturation exists. If we want to understand the dynamics of development we probably must first examine what underlies it on the biological side, which does not mean that other factors will not be considered as they deserve. I do not want either to give you the impression that in my mind scientific research is more important than clinical work. I have said already that our therapeutic efficiency in this field was limited; our only logical hope to reduce the number and severity of cases of cerebral palsy and allied conditions lies in rational scientific prevention.

Method of Investigation

What means of investigation can we use if we are to understand the intimate "why and how" of maturation and its pathological deviations? We can use clinical methods. I want to remind you here of the work of a senior Minkowski of Zurich, of Davenport Hooker on foetuses, of André Thomas and Madame Saint-Anne Dargassies on very young prematures; Minkowski Jr. will show you a very remarkable film on this subject. All this has nothing to do with a purely descriptive, botanical semeiology; the type of motility seen at a given age gives valuable clues concerning the level of neural organisation. Histology, mainly with Leroy-Connell, adds to this knowledge, and another approach is now possible with the help of our EEG colleagues. Here I will quote the names of Smith, Lindsley, Hill, and, in France, Fischgold's team and mainly Madame Dreyfus-Brisac and her co-workers; their findings will be presented to you by Minkowski. All these investigations—clinical, histological and electroencephalographic — show that the process of maturation is not a linear one; there are jumps, followed by slower periods of organisation and integration of progress.

Another channel of investigation is developing fast—I mean, biochemistry of the developing nervous system. I have neither the knowledge nor the time to summarise the work accomplished in this field; an important symposium dealing with these aspects was held here in Oxford in 1954 and its proceedings have been edited by Waelsche. I will simply mention the importance of cerebral lipids, proved by the very process of myelinisation and the existence of cerebral dyslipidosis. Congenital galactosæmia teaches us that carbohydrates are equally important, and phenylketonuria reminds us that we have also to deal with aminoacid metabolism. Several authorities, and among them the Russians, have stressed the importance of enzymatic systems, mainly cytochrome-oxidase and succinic-acid-dehydrogenase. The problem is linked with that of the utilisation of oxygen. Another problem arising is that of the "B.B.B.", or blood-brain barrier; but this is obscure even in the mature organism.

More recently the influence of vitamins has been studied; Giroud has shown that vitamin A can play an important part in the determination of congenital malformations. The role of endocrine glands is going to be reviewed at the present meeting. And finally we have to consider the genes. Some of them obviously carry an abnormal trait, and others might bring a predisposition, a susceptibility to external

factors; this is perhaps the story of mongolism.

There are obviously tremendous gaps in our knowledge: we do not know in many instances what goes wrong with the maternal organism. This problem is still complicated by the fact that the mother's condition can be harmless to her and still damage the fœtal brain; this is what happens in rubella, toxoplamosis and blood incompatibility, the three main discoveries in our field in the last twenty years. And, finally, our knowledge of the physiology of the placenta is still incomplete. We know more about what happens during delivery; quantitative data concerning the degree of anoxia as they have been studied by the Minkowski group will be very valuable. Factors influencing vacular resistance in the premature and the newborn must be considered too, and the same applies to the possible damage caused by bilirubin in excess.

In this field our present knowledge enables us to avoid or to prevent some of the morbid conditions we have to deal with. Congenital cerebral syphilis has become a rarity; up to a point we can help the Rh-positive child of an Rh-negative mother to escape from the threat of cerebral involvement; we can and must minimise in all cases the damage caused by anoxia, intracranial hæmorrhage or œdema. A tremendous hope arises in connection with what we know about the pathology of congenital galactosæmia and phenylketonuria; it seems that a combination of early diagnosis and proper diet can save the future of such children. But there remains the large number of unknown, misunderstood conditions. I have already stressed the importance of an early diagnosis. But I want to emphasize the importance of an *accurate* diagnosis.

Human Relations

Now let us leave the field of science for that of human relations. We must admit that modern psychology started with Freud. A few other names might be quoted, those of Melanie Klein, Anna Freud and René Spitz being the most important. There is a striking difference between the laboratory animal, from the study of which we have gained a great part of our scientific knowledge, and the human infant, not only because the state of preparedness for life at birth varies from one species to the other, but chiefly because in the human infant we have to deal with specific human emotional problems. The crippled child is not only a neurological or biological case, he is his mother's child—he is the object of long expectations, of dreams and fantasies. If you tell this mother in an abrupt way that her child is going to be a cripple, an idiot, that he will never be normal, this will determine a deep depressive state. Personally I think that we have a duty to tell the truth, but we must be sure that it is truth and we must say it with the greatest solicitude.

Another point emerges from this—that of classification. I think classification is useful, but not in the sense of superficial clarity, as in a botanical classification. We have to know what a condition means in terms of the future; that is the main problem for the parents. Prognosis and possible therapy are more important for them than diagnosis. To appreciate the prognosis of a case of cerebral palsy two factors are important: motor abilities and psychological potentialities. There is no rehabilitation if the child does not cooperate, if he is not willing to make progress, to gain, to get closer to the parents with whom he obviously identifies himself. And this stresses again the importance of psychological factors.

Another problem arises in connection with the diagnosis of cerebral palsy. The mother when she learns the truth is often impatient to do something, to start at once with a programme of technical rehabilitation. But we know that the young child is not prepared to live without his mother; if we put him into an institution

we add to the somatic impairment the noxious effects of what Spitz has called "hospitalism". During the first few years there cannot be a better therapist than the mother herself.

Towards Rehabilitation

I will not deal with the purely technical aspects of rehabilitation; there are enough competent people in the audience to do it. I will confine myself to some specific aspects of this new type of Medicine. The part devoted to non-medical technicians is growing; the doctor is no longer an omniscient magister, he is becoming more and more humane, he is the leader of the rehabilitation team. But other members have probably a more important role; emotional factors are so prominent that the really active part is usually played by women, because they are more able to become a kind of mother substitute. But there are also technical aspects in rehabilitation. We have here several specialists in orthopædics, electrical muscular testing, and even neurosurgery who can tell you better than I what is to be expected from their knowledge. The growing science of neuropharmacology provides us with new antispasmodics and tranquillizers; it might be worth while to discuss this topic briefly during this meeting.

And, finally, the impact of epilepsy should, to my mind, be dealt with. First of all the association of epilepsy with cerebral palsy aggravates considerably the inherent psychological difficulties. We briefly discussed this problem at the IVth International Congress of Child Psychiatry held in Lisbon last June. The second point is that, according to the work of Scholz and Morel, among others, it seems that recurring epileptic activity aggravates cerebral lesions, mainly in the area of Ammon's horn, thus further impairing intellectual development.

Once we are faced with a "cerebral palsy" child we cannot hope to correct or eradicate an already established lesion; but we have to get the optimum result for the structures that have been spared. This is why we have to deal with different kinds of specific learning difficulties. Some of these children will have a distorted perception of space, time or rhythm, others will be world-blind or dysorthographic or dysarithmetic. Very often, the body image, built in the normal child through normal movements, will be impaired. And at the same time the crippled child will soon realise that he looks different from other children. Lay people often use the term "inferiority complex"; the "cerebral palsy" child has every reason to develop it and that is why psychotherapy is a very important part of this work of rehabilitation and specialised teaching.

Summing up, there was a time, not too far remote, when Child Neurology was an arid and hopeless subject. Now it is oriented towards research and rehabilitation; it is becoming more and more dynamic. It is still a hard fight, but each success means that a human being has been enabled to carry on a normal existence.

THE CHARACTERISTICS OF SPASTICITY IN CHILDREN*

RONALD MAC KEITH, M.D., F.R.C.P.

From the Department of Child Health, Guy's Hospital, London.

SPASTICITY is a special type of hypertonus, hypertonus being increased resistance of the muscles to passive movement at a joint. Spasticity presents in four ways:

(1) On passive movement of a joint, there is in one direction a resistance which, when pressure is steadily increased to an adequate level, yields slowly and steadily.

(2) On passive movement of a joint there is, in one direction, a resistance which, when the muscle tension reaches an adequate level or a certain muscle length is exceeded, gives way suddenly. This is sometimes referred to as a "give".

(3) A sudden resistance of the muscles on more or less quick passive movement. This is sometimes referred to as a "catch".

(4) Hyperreflexia. This "is only of significance in relation to other evidence or when one-sided." (Denny Brown, 1957, in his *Handbook of Neurological Examination*).

If any of these four is present, there is spasticity.

Denny Brown (loc. cit.) says of spasticity "It is constant from hour to hour, and varies from day to day." This is not true of the spastic child, whose spasticity can vary from minute to minute or second to second.† It will be greater if he is cold. It is much affected by the emotional state of the child. If he is frightened or startled, the spasticity may be more obvious. The child's posture will affect the amount of hypertonus in various limbs; it may vary according to whether he is prone or supine; or according to the position of his head relative to his trunk.

The distribution of spasticity varies with age. For example, a spastic diplegic at one year may have extension at the knees; at 15 they may be in flexion. An operation for lengthening the tendo achilles abolishes spasticity from the plantar flexors of the ankle. After immobilisation in plaster for a time — for example, in the treatment of congenital dislocation of the hip—spasticity may disappear and be replaced by hypotonia, which is even more disabling than the spasticity.

†Note by Recorder: The use of this quotation is unfair. Spasticity, even in children, is constant provided it is examined under comparable conditions both of external and internal environment. This of course applies to all reflexes, including stretch reflexes.—G. Rushworth.

*Read at the N.S.S. International Study Group on Child Neurology and Cerebral Palsy, Oxford, Sept. 21-27, 1958.

MUSCLE TONE AND POSTURE IN INFANCY*

T. T. S. INGRAM, M.B. (Edin), M.R.C.P.E., D.C.H.
From the Department of Child Life and Health, University of Edinburgh

THE AGE at which cerebral palsy is first diagnosed has fallen in Edinburgh from an average in 1952 of 3 years and 2 months to 10 months in 1957-58. This improvement in diagnosis may be attributed largely to the readier recognition of the early manifestations of the condition, and the realisation that the clinical picture presented by cerebral palsy in infancy may be very different from that shown by the patient a few years later.

It is the purpose of the present paper to describe the changes in posture, control of posture and muscle tone which occur during maturation in normal infants, and compare them to those observed in patients with bilateral cerebral palsy.

Theoretical Considerations

Though neither "tonus" nor "posture" can be easily defined, they are so closely related in terms of the underlying nervous activity which produces them and in their functions that it is impossible to consider one without the other. Muscle tone and posture are not isolated physiological phenomena. The maintenance of posture is the function of muscle tone. The corollary is that muscle tone will be greatly influenced by changes in posture. Jakob's description (1925) of posture as a state of arrested movement emphasises the active function of muscle tone in maintaining posture in most states of waking immobility. Since muscle tone is so altered by changes in posture, the classification of cerebral palsy on the basis of states of muscle tone may be unsound, especially in very young patients.

In general, postural changes effect a greater alteration of muscle tone when the nervous system is comparatively simple than when it is highly organised. The reflex activity of the spinal cord is modified by the integrating and partially inhibitory activities of the higher centres in the brain stem. The alterations in motor activity which are due to centres in the cord and brain stem are modified by those in the basal ganglia; and in turn those of the basal ganglia, brain stem and cord complex are partially inhibited and integrated by centres of nervous activity in the cerebrum. The decreasing effect of postural changes on muscle tonus as higher centres of nervous activity come into play was well shown by the classical work of Sherrington on various spinal and decorticate animal preparations (Sherrington 1947, Walsh 1957, Bard and Macht 1958).

In the human infant, changes of posture produce less alteration of muscle tone as the child matures. Many of the newborn's primitive motor reflex patterns are mediated at a cord or brain stem level. As higher centres of nervous activity come into play these patterns are progressively inhibited and integrated. Alterations in posture which once resulted in gross changes in muscle tone no longer do so. The primitive reflexes which are charac-

*The substance of this paper was given at the National Spastics Society International Study Group on Child Neurology and Cerebral Palsy, Oxford, Sept. 21-27, 1958.

teristic of infancy disappear in a consistent and predictable order as the child matures and the nervous system becomes more complex and highly organised. Thus their presence or absence in an individual child are fair indications of the level of motor maturation which he has reached. For example, the prognostic significance of the disappearance or persistence of the grasp reflex, symmetrical and asymmetrical neck reflexes, body righting reflexes and falling reflexes is great. The elicitation and interpretation of motor patterns and reflexes characteristic of the various levels of neurological maturity in infancy amounts to a system of developmental neurological diagnosis (Gesell 1940, Thomas and Sainte-Anne-Dargassies 1952, Koupernik 1954). The relationship of the various patterns of motor behaviour to the underlying structure of the nervous system is still largely unknown.

The transient alterations in muscle tone which result from changes in posture are superimposed on a state of persistent tonus present during waking hours. Though this persistent tonus varies with individuals, emotions, fatigue and many other factors, the distribution in different muscle groups of relative persistent hypotonia and hypertonia tends to be characteristic for each level of neurological maturation. The successive stages of progressively more complex neurological organisation which occur as higher nervous centres come into action are each manifest by specific patterns of motor behaviour and characteristic distributions and intensities of muscle tone. These states of muscle tone tend to be continuous, though they will be transiently altered by postural changes (McGraw 1943, Gesell 1954).

Fig. 1 (a) and (b).—The characteristic resting posture of the newborn baby. The limbs tend to be semiflexed.

Posture and Tone in the Normal Infant

At birth the child is usually postureless and flaccid, but with its first breath posture and tone in the muscles become apparent. The posture is one of semiflexion. Tonus is greater in flexor than extensor muscles. The child seems to like to lie on one side with the limbs in a position of modified flexion—"a modified fœtal position". The semiflexed posture, in which the head is semiflexed on the trunk, the primary spinal curve is marked, and all four limbs are in semiflexion, is the position of comfort. The infant tends to return to it after active or passive movement (fig. 1). This is most clearly seen when the Motor reflex is elicited. This is an "alarm" or "startle" response. A sudden movement or in some cases even a loud noise causes the child to make a momentary involuntary extension of the

neck, trunk and limbs which is followed by a more gradual return to the position of generalised semiflexion which has been described. When the tonic neck reflex is elicited at this stage it is characteristic that the flexor movements of the limbs are greater in range and stronger than the extensor. The period during which the resting position of the waking child is one of semiflexion may be described as the first flexor stage of infancy.

The child's resting position becomes less markedly flexed in the weeks after delivery. At about the time when the child first holds up its head unsupported, momentarily, the flexor (second) stage of the Moro reflex diminishes in intensity. Sudden movements of the child now result in the neck, trunk and limbs being extended as before, but the position to which the child returns is no longer one of such extreme semiflexion.

Some normal babies aged between 4 and 12 weeks show such a marked tendency to transient extension of the trunk and limbs when startled, without an immediate return to the semiflexed position which they showed a few weeks earlier, that they are sent for medical consultation. The common complaint is of "sudden back arching", "going stiff" or by the more sophisticated of "extensor jerks", which occur whenever the child is alarmed.

The tendency to persist in extensor postures for a period after involuntary or active movement is an indication that the first flexor stage is coming to an end. As these changes in resting posture take place there is a relative decrease in flexor tonus and an increase in extensor tonus in the limbs and trunk.

By the time the child can hold up the head independently and steadily, transient increases of extensor tone are less marked. The extensor phase of the Moro reflex diminishes in intensity. The extensor movements of the limbs are more marked and powerful than the flexor when the tonic neck reflex is elicited. But the reflex becomes less easy to demonstrate towards the end of this stage. At the same time the position of comfort has changed considerably, so that in the waking state the neck, trunk and limbs are much more extended than they used to be. (During deep sleep, positions of semiflexion persist into early childhood). A phase characterised by transient increases in extensor tonus is succeeded by a phase in which there is persistent extensor, or antigravity, hypertonus. The presence of persistent increase of tone in the extensor muscles of the trunk and limbs becomes progressively apparent in the course of a few weeks. Passive mobility of the limbs is decreased and a minimal degree of "lead pipe" rigidity may even be appreciated in the limbs on examination. These features are characteristic of what may be termed the "first extensor stage" of infancy.

While the tendency to extensor positions of the limbs and a relative increase of tone in the antigravity muscles is persistent in this stage, the degree of antigravity hypertonus may easily be increased reflexly by a number of manœuvres. Sudden passive extension of the head, pressure on the soles of the feet, putting the child into the erect position will all tend to produce a marked increase of tone in the extensor muscles of the trunk and limbs. This is most manifest in most children between the ages of 4 and 6 months, when reflex standing becomes possible (fig. 2). If the child is placed in the erect position with its bare feet on the floor and the head tilted back, tone in the antigravity muscles is such that it can bear its own weight and requires only minimal support to the hands to prevent tilting in one piece forwards, backwards or to one side (fig. 2). This phenomenon may be regarded as an extreme expression of the alterations in the distribution of muscle tone which occur in the first extensor stage.

The first extensor stage does not last for more than a few weeks. Reflex stand-

ing is easily elicited for only 6 to 8 weeks in most children; it becomes more difficult to produce perceptible increase in anti-gravity hypertonus by the manœuvres which have been described. Instead of extending the head and limbs and trunk when held inverted by the feet as he did in the extensor stage, the child now flexes the neck and back and limbs when placed in this position. He may even look as if he is trying to climb his legs. At this stage voluntary flexion of the trunk becomes possible and the child gradually becomes able to sit without support. On examination a gradual increase of tone in the flexor muscles is perceptible and may be sufficient to give quite brisk stretch responses and lively biceps and supinator jerks. The tone in the extensor muscles of the limbs and trunk is noticeably less than that in the flexor groups. These changes are characteristic of the second flexor stage.

Fig. 2.—"Reflex standing" in a normal baby aged 4 months. With minimal support the child can maintain the erect position when the head is extended and there is pressure on the soles of the feet.

Fig. 3.—Crawling with reciprocal limb movements in a child aged ten months. The upper limbs are extended at the elbows. Though the knees are still flexed they are lifted from the ground during crawling movements. These patterns are characteristic of the early second extensor stage

The second flexor stage is less well defined than previous stages, largely because of the increased amount of highly co-ordinated voluntary motor activity which is evident during it. It merges almost imperceptibly towards the end of the first year into the second extensor stage. This is characterised by the gradual assumption of the erect position. From crawling on his forearms, knees and the fronts of his feet the child graduates to crawling on his hands and feet with the limbs extended (fig. 3). He will attempt to climb up his cot side with lower limbs and back extended before he can stand without support. When placed on his abdomen he will tend to extend his back and neck instead of flexing them, and will reach upwards, often attempting to climb on any available support. When held upside down by the feet the head will extend and he will look at the floor. At the same time he arches his back and extends the limbs, quite often placing the hands to the ground. If he is held upright and suddenly tilted forwards, he will extend the upper limbs and stiffen the trunk by increasing the tone of the antigravity muscles, as if to protect his head from trauma (Magnus 1924). The effect of postural changes in altering muscle tone are much slighter than they were in previous stages. The major alteration in continuous muscle tone is a diminution in the degree of flexor hypertonus and some reduction in the

Fig. 4.— Walking with little support, aged 11 months. The achievement of unsupported walking is the major motor achievement of the second extensor stage.

activity of the biceps and supinator jerks, and (rather inexplicably) the knee jerks The main motor achievement of the second extensor stage is the assumption of the erect position. The child stands, first with support, then without support, and after a little time walks (fig. 4).

Tone and Posture in Cerebral Palsy

The changes in posture and alteration in muscle tone which occur in many cases of cerebral palsy may be interpreted in terms of the various stages of neuromuscular maturation observed in the normal infant. It is possible to consider the clinical findings in some forms of cerebral palsy as the result of retarded neuromuscular maturation, or the failure to develop centres of higher nervous integration.

Thus in very severely affected microcephalic children, the resting position of semiflexion may never be lost. The Moro reflex may persist in both its extensor and flexor phases. Many of these children remain somewhat hypotonic for many years. They may have jerking attacks in which fragments of the Moro reflex, either extensor or flexor in type, may be apparent. Such patients rarely mature beyond the stage of achieving incomplete control of head posture (Yannet and Horton 1952).

Diplegia

In diplegia a retarded but orderly progression of neuromuscular maturation through the stages observed in the normal child may be discerned (Ingram 1955). The manifestations are gross and uninhibited, but they are unmistakable to anyone interested in the motor behaviour of normal infants. Though caricatures of the phenomena observed in babies following a normal pattern of development, they are all the more striking and recognisable on this account. Their significance has been largely ignored (Illingworth 1958).

The semi-flexed position of the normal child persists for a much longer period in the moderately severely or severely affected diplegic patient. The child may remain relatively immobile for months instead of weeks. At this stage the diplegic children show only the normal patterns of behaviour of the newborn for rather longer than the trained observer might expect. On physical examination he would be unlikely to diagnose cerebral palsy and more likely than not would be wrong if he did so. The period for which the patient remains hypotonic and semiflexed when resting depends greatly on the extent and severity of his diplegia, and presumably on the effect of the underlying anatomical defect on the nervous system.

The first definite abnormality observed by the parents is commonly the appearance of sudden transient extension of the head and trunk, and often the limbs, when they handle the diplegic infant, often when bathing or dressing him. The position is characterised by hyperextension of the head and neck and trunk, adduction of the shoulders, extension of the elbows, pronation of the forearms, flexion of the wrists and fingers. The hips are extended

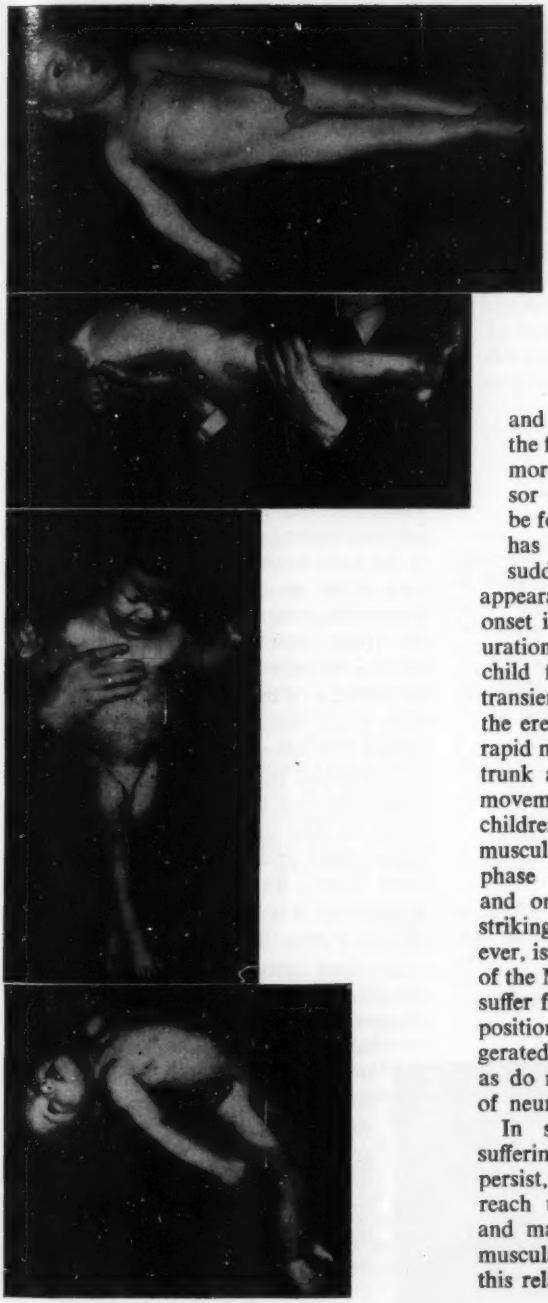

Fig. 5 (a), (b), (c) and (d).—Characteristic postures of a diplegic child in the dystonic stage. (a) Boy aged 3½ years. (b) Boy aged 11 months with head thrown backwards. (c) Same child as in (b) with head flexed in the erect position. (d) Same child as in (b) and (c) aged 20 months with head gently extended and turned to show tonic neck reflex and less marked symmetrical neck reflex.

and adducted, the knees extended and the feet plantar flexed and adducted. On more detailed questioning these "extensor spasms" or "dystonic attacks" will be found to occur when the child's head has been allowed to extend relatively suddenly. The chronological time of its appearance varies greatly, but at its time of onset in the scale of neuromuscular maturation is consistently about the time the child first begins to hold the head up transiently when passively maintained in the erect position. The similarity of these rapid movements of extension of the head, trunk and limbs to the sudden extensor movements observed in some normal children at a similar stage of neuromuscular development, after the flexor phase of the Moro reflex has declined and only the extensor phase is left, is striking. This extensor movement, however, is not identical to the extensor phase of the Moro response (fig. 5). Patients who suffer from diplegia show much the same positions of comfort and similar, if exaggerated, responses to changes of posture as do normal children at the same stage of neuromuscular maturation.

In some severely affected children suffering from diplegia dystonic attacks persist, and though some of them may reach the stage of sitting with support and maintaining the head upright transmuscular maturation becomes arrested at this relatively primitive stage.

Fig. 6. — Characteristic "reflex standing" posture in the rigid stage of diplegia.

Fig. 7.—Boy suffering from diplegia, aged 6 years, showing posture characteristic of the transient phase between the rigid and spastic stage of the condition.

Fig. 8.—Same child as in Fig. 7, showing that sudden extension of the neck still produces an increase in antigravity hypertonus.

The majority of patients with diplegia advance beyond this stage. Transient involuntary extensor movements (dystonic attacks) become less frequent and less severe. As they decline there is a gradual increase in continuous extensor, or antigravity, hypertonus. The resting position changes from one of generalised semiflexion to one of modified extension in neck, trunk and limb muscles. As in the normal child at this stage of neuromuscular maturation increased tone in the hamstrings and adductor muscles of the thighs is apparent. "Reflex standing" may be possible, and it is often necessary to point out to parents and therapists that this phenomenon does not mean that the patients are approaching the stage of independent standing and walking (fig. 6). The point may be emphasised to both by pointing out to them that the child has so far been unable to sit without support, even when the knees have been flexed, since sitting requires a flexor attitude of the trunk which the child cannot yet achieve voluntarily. On examination it is the stiffness of the limbs which is striking. Here is no "minimal degree of lead pipe rigidity" observed in the normal child at an equivalent stage of neuromuscular development. The limbs are stiff and difficult either to flex or extend. Little's phrase "generalised rigidity" is appropriate in these cases. But though hypertonus is generalised it is most evident in the antigravity groups (fig. 6).

In some patients neuromuscular development ceases at this stage of generalised rigidity with predominant extensor hypertonus. The patients are stiff and immobile. Extension of the neck, the erect position, and pressure on the soles of the feet all increase antigravity tonus still further. The tonic neck reflexes are very marked, but true spasticity, as demonstrated by increased stretch responses and consistently increased tendon jerks, is absent. These patients do not walk, use

their thumbs or release their grasp, and they cannot sit without support. These are measures of their neuromuscular immaturity.

In the majority of patients, however, there is further neuromuscular maturation. This is demonstrated by a gradual increase of flexor tonus and a decrease of extensor, or antigravity, hypertonus after a period of weeks or months, depending on the rate of development. However, maturation may cease at any stage, and some patients show persistent extensor hypertonus and resting positions of predominant extension even when flexor hypertonus (spasticity) may be demonstrated in the limbs (fig. 7). These children often trouble their therapists greatly by persistent "extensor thrust" whenever they are placed in the erect position. The additonal reflex stimulus of the erect position and pressure on the soles of the feet still results in an increase of antigravity hypertonus, as in the normal child at this stage. There is an uncontrollable involuntary contraction of the extensor muscles of the limbs and trunk. The head is thrown back by the contraction of the extensor muscles of the neck (fig. 8). "Extensor thrust" is easily explicable if the sequence of events in normal development is remembered, but it is often regarded as a manifestation of the child's "cussedness" by therapists and doctors who feel that "a child who can stand should do better."

Most patients suffering from diplegia advance beyond the stage of obvious persistent antigravity hypertonus. In them the degree of hypertonus in the flexor muscles of the trunk, neck and limbs gradually increases and the hypertonus in extensor muscle groups diminishes. Stretch reflexes in the flexor muscles of the limbs become exaggerated. The biceps, supinator, knee and ankle jerks become brisker. Ankle clonus, and less often patellar clonus, may be elicited. The plantar responses assume a convincing extensor pattern bilaterally. The patient becomes truly spastic and is likely to develop flexor contractures in spite of what his therapists and orthopædic surgeons do to prevent him (fig. 9).

Dyskinesia

The early development of children who suffer from an excess of involuntary movement as their predominant disability ("athetoids", "choreo-athetoids", "dyskinetics") is rather similar to that shown by patients with diplegia. These patients are commonly somewhat immobile and hypotonic during the first weeks of life, though they are not strikingly enough abnormal to worry their parents. The first symptom which worries many of them is the tendency of their children to extend their necks, trunks and limbs suddenly while being handled, and especially while being bathed or dressed. The posture the patients assume at this stage is exactly that described for children suffering from diplegia. There is hyperextension of the neck and trunk, adduction at the should-

Fig. 9.—Characteristic flexed posture in the spastic stage of diplegia shown by a child aged 6 years. At this stage sudden extension of the head has relatively little effect on posture.

(a)

(b)

(c)

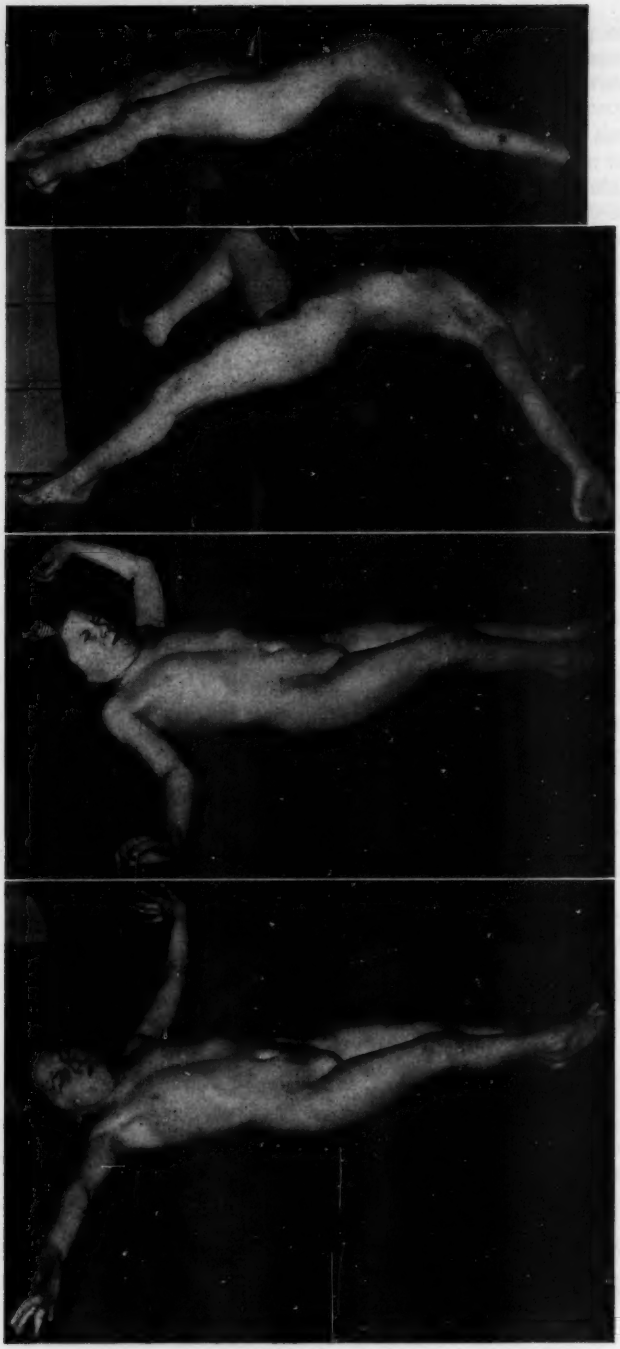

Fig. 10 (*a*), (*b*) *and* (*c*).—*Girl aged* 8 *years with severe dyskinesia. The involuntary movements are predominantly dystonic in type.* (*a*) *Symmetrical neck reflex, showing posture resulting from sudden extension of the head.* (*b*) *Tonic neck reflex.* (*c*) *Residual fragments of the Moro reflex.*

ers, extension of the elbows, pronation of the forearms, and flexion of the wrists and fingers, commonly over the adducted thumbs. The hips are adducted, the knees extended and the feet plantar flexed and internally rotated. The position is one of opisthotonos. The sudden extensor movement which produces them may be termed a "dystonic attack". Thus in children suffering from very severe "dyskinesia" there may be no advance from this stage of neuromuscular development. Extensor spasms of the trunk and limbs persist, the ability to reach is not developed, and ability to hold up the head steadily is not achieved. On examination the Moro reflex can usually be elicited, and the tonic reflexes are marked, being associated with particularly powerful extensor movements of the limbs (fig. 10).

The majority of patients suffering from dyskinesia advance beyond this stage of neuromuscular maturation. The mass involuntary extension of the neck, trunk and limbs becomes less violent. At about the time voluntary control of the head is achieved, passively produced changes in posture result in less severe extensor hypertonus, though some "extensor thrust" persists in patients when they are placed in the erect attitude. Voluntary activity of the limbs is interrupted by involuntary movements characterised by transient extensor hypertonus, or "tension" as Phelps terms it. Some of the involuntary movements of the limbs in many cases of choreoathetosis may be regarded as fragments of the previously more generalised, massive involuntary extension of the neck, trunk and limbs. The relatively slow involuntary movements maximal in the proximal parts of the limbs (dystonia*) are the most easily recognisable fragments of the more generalised mass movement of extension previously present.

As further neuromuscular maturation occurs, the transient extensor hypertonus becomes less marked in the limbs as well as in the trunk. But involuntary movements consistently diminish in severity in a cephalocaudal direction. Head posture improves before control of the trunk is achieved. The control of voluntary movements in the proximal joints of the limbs is evident before the hands can be used with accuracy. In this the pattern of functional maturation is similar to that observed in the normal child.

Conclusions

It is possible to interpret many of the clinical manifestations of early bilateral cerebral palsy in terms of the characteristic neurological behaviour patterns of the normal child. The same stages of neuromuscular development may be discerned, though in the patient with cerebral palsy the rate of maturation is much slower, and in many cases it ceases at a relatively primitive stage.

The postural responses and alterations of muscle tone which occur in patients at the various stages of neuromuscular maturation are similar in character to those observed in the normal child at equivalent stages, but they are grossly exaggerated in degree, because the normal inhibitory function of higher nervous centres is incompletely developed.

References

Bard, P. and Macht, M.B. (1958) in **Neurological Basis of Behaviour.** London: Churchill.

Gesell, A. (1940) **The First Five Years of Life.** London: Methuen.

(1954) in **Manual of Child Psychology.** Edited by L. Carmichael. 2nd Ed. London: Chapman & Hall.

Illingworth, R. S. (1958) **Recent Advances in Cerebral Palsy.** London: Churchill.

Ingram, T. T. S. (1955) "Early Manifestation and Course of Diplegia in Childhood". **Arch. Dis. Childh., 30,** 244.

Jakob, A. (1925) "The Anatomy, Clinical Syndromes and Physiology of the Extra-pyramidal System". **Arch. Neurol. Pshychiat., 13,** 596.

Koupernik, C. (1954) **Développement Psycho-moteur du Premier Age.** Paris: Presses Universitaires de France.

Magnus, R. (1924) **Korperstellung.** Berlin: Springer.

MacGraw, M. G. (1943) **The Neuro-muscular Maturation of the Human Infant.** New York: Columbia University Press.

Sherrington, C. (1947) **The Integrative Action of the Nervous System.** Cambridge University Press.

Thomas, A. and Dargassies, Sainte-Anne (1952) **Etudes Neurologiques sur le Nouveau-Né et le Jeune Nourrisson.** Paris: Masson & Perrin.

Walsh, E. G. (1957) **Physiology of the Nervous System.** London: Longman.

Yanet, H. and Horton, F. (1952) "Hypotonic Cerebral Palsy in Mental Defectives". **Pediatrics, 9,** 204.

*For some of us, the use of the word dystonia for movements is undesirable, for dystonia is an abnormality of tone, to be detected by handling the limb, rather than by looking at it.—Ed. **C.P. Bulletin.**

Educational Disabilities of Cerebral Palsied Children attending Local Education Authority Schools*

R. V. SAUNDERS, M.A., B.ED.
From the Bristol Child Guidance Clinic.

I SHOULD explain that I am responsible for the psychological work in the Bristol school psychological service, and in the Bristol L.E.A. Child Guidance Clinic. I have also done practically all the psychological assessment work in Dr. Grace Woods' Cerebral Palsy Assessment Clinic at Bristol Children's Hospital, where we have now seen over 500 children mostly from the hospital clinical area. So far as the Bristol L.E.A. is concerned, I not only make the initial psychological assessments of the Bristol children attending the clinic but also follow them up in the Bristol school system.

Educable Bristol cerebral palsied children are placed in a variety of ways:

(a) In our day spastics school.
(b) In ordinary schools.
(c) In schools for educationally subnormal pupils.
(d) In our school for physically handicapped pupils.
(e) A few in independent day or residential special schools.

I propose to mention the difficulties of children attending our own Bristol schools.

The Day Spastics School

This is a small school for 35 children which is just beginning to develop a post-primary stage for children over 11 years old. I believe this to be an exceptionally good school with a Head who can take most problems in her stride. But we have found that certain types of C.P. children do not fit in well with the smooth operation of the school. Recently we admitted a small girl who behaved very well and showed a good response at the clinic, where she had the undivided attention of the adults. On her admission to the school, however, it soon emerged that she was not prepared to share the attention of the staff with the other children, and she asserted her demands through violent temper tantrums. Since she was quite mobile and was also inclined to interfere with the postures of the other children, few of whom were able to take evasive action, she produced states of tension in the pupils which were a serious threat to the work of the school. We had therefore to arrange for her transfer after only one term. The point which concerned us most was that the clinic attendance and assessment did not, and could not, I think, be expected to bring out the difficulties which emerged when she began to attend school. A comprehensive social history might have done so. Observation of the child in her own home would certainly have done so in this case.

Transition from Spastics School to Other Schools

We have learnt to approach this transition with very great caution. A child who seems ready to leave our day spastics school is not ready for the robust life of an ordinary school without careful preparation. All sorts of unexpected pressures materialise. The child is no longer trans-

*Read at the N.S.S. International Study Group on Child Neurology and Cerebral Palsy, Oxford, September, 1948.

ported to school, he may have to stand in dinner queues, he will not have any afternoon rest, and, most important of all, he will have to share his teacher with some 40 other children when previously there were only 8 to 10 in his group.

In these circumstances, it is not surprising to find that children transferred to ordinary schools may fail to make the same educational progress as before; those of anxious disposition may become more anxious and depressed, while some even develop hysterical symptoms. A great deal depends on how supporting and reassuring parents can be. Those with a strong wish for the "normality" of their child can produce a drive for educational attainment, especially in reading, which creates an anxious, sometimes obsessional, child who adjusts poorly to the social life of the school. On the other hand, some parents are too protecting and undemanding and we get either the euphoric child who will not apply himself to any kind of educational achievement, or the child who is too timid to try.

C.P. Children in Ordinary Schools

The general impression obtained from a survey of educational progress in relation to the ability of these children is that C.P. children of less than average ability tend to be educationally retarded, and that it is only among those of high average or better ability that one finds reasonably good educational progress and adjustment. The C.P. child of less than average ability appears likely to make an even poorer educational adjustment in the ordinary school setting if the EEG picture is poor.

What are the best criteria for determining the suitability of C.P. children for attending the ordinary day-school? To judge by Bristol experience, a single criterion of minimal locomotor handicap is not enough. It seems that one must consider: (*a*) motor capacity; (*b*) E.E.G.; (*c*) possible auditory and/or speech defects; (*d*) possible spatial and other perception difficulties, which may exist alongside a minimal physical handicap; (*e*) intellectual capacity; (*f*) temperamental qualities; and (*g*) social adjustment both to adults and to other children.

In short, one must make a comprehensive case-study of each child to be reasonably sure of avoiding misplacements. In my view, such a study is done infinitely better by a team than by one individual, and the basic team should include a pædiatrician, a psychologst and a a social worker with facilities not only for interviewing the child's parents at the clinic but also for visiting them in their homes, to see the child in its own setting and assess the family atmosphere. This last is the item which we in Bristol feel to be most wanting in our organisation.

From a quick survey of our records the following points appear most worthy of mention.

Among our Bristol cases, right hemiplegics appear to be the group most normally distributed on the ability scale. (Dr. Woods has commented on their superiority to left hemiplegics in her book, "*Cerebral Palsy in Childhood*").

Athetoids probably tend to be underrated by intelligence tests. This appears to be due to two considerations: underrating on verbal scales owing to speech handicaps, and also possibly to hearing defects; and underrating on performance scales because of poor manipulative ability.

Many C.P. children assessed as of dull ability tend to find their way into special classes or schools for educationally subnormal children. My present impression is that this is due not so much to specific defects as to a more general slowness of response in comparison with the normal child in the ordinary school situation. In saying this of C.P. children as a whole, it must be remembered that particular types of these children may tend to exhibit specific receptive defects more than others.

Over the past few years I have worked

with retarded C.P. children attending ordinary schools on an individual coaching basis. The response to one half-hour's individual teaching per week has been encouraging, though it is sometimes necessary to wait for as long as 18 months to obtain substantial results. This experience has convinced me that remedial work in small adjustment groups could usefully be tried with retarded C.P. children who attend ordinary schools. The teachers who have such children in their groups should have contact with the local spastics school or unit, so as to understand the learning difficulties with which they may have to deal.

In discussing or assessing intelligence, I find it valuable to bear in mind the distinction which Hebb makes between Intelligences A, B and C. *Intelligence A* is "an innate potential, the capacity for development, a fully innate property that amounts to the possession of a good brain and a good neural metabolism". *Intelligence B* is "the functioning of a brain in which development has gone on in a 'completely stimulating environment', remembering that some environments are more facilitating and stimulating than others". To these one should add *Intelligence C*, the individual's intelligence-test score—i.e., the result of an attempt to assess Intelligence B. It is extremely useful to keep these distinctions in mind in assessing any child, but more particularly in assessing handicapped children.

A psychologist should not be regarded simply as "someone who does IQ's". His function is to "assess children", and in the process he does three things:

(i) Obtains a test result in the form of a standard score, or set of scores, or an IQ rating.

(ii) Records the child's reactions to the test and the test situation. This is qualitative information of clinical value, and can be obtained even when a quantitative result is not forthcoming.

(iii) Evaluates the child's reaction to himself as a person. This also is information of clinical value, which, together with the previous point, may have a considerable effect on the amount of reliance to be placed on the quantitative result.

In conclusion, I would like to underline Dr. Koupernik's advice—that we must always consider the child *as a person*, and whatever we do for him must have regard to the unique dynamics of his individual life situation.

SUMMARY OF DISCUSSION ON MR. SAUNDERS' PAPER

Right and Left Hemiplegics

The comparative performance of these two groups provoked considerable discussion. Dr. Grace Woods pointed out that the Bristol hemiplegics are at present being defined by their movement handicaps. She was not attempting to explain why the right hemiplegics showed themselves superior—it had happened that way. An independent psychological study by Dr. Weddell in investigating perceptual problems in a C.P. population had shown a superiority of right over left hemiplegics, so that the superiority showed itself both in intelligence-test and perceptual-test performance.

Among the comments on these findings, perhaps the most valuable was that we are not strictly entitled "to identify the hemiplegics with a definitely one-sided lesion". Other comments touched on ætiology—e.g., that mental deficiency is commoner in congenital than in acquired hemiplegia, and that there are more left than right congenital hemiplegics.

Team Work

Several speakers endorsed the need for, and value of, team-work. A distinction was made between the basic team, who did the routine diagnostic work on each new case, and the considerably larger array of specialised workers who could

be called in at need, but whom it would be uneconomical to have in on every case. Dr. E. Ellis described his interesting unit which admits children with their mothers for a week, allowing time for assessment of the child's difficulties.

Sheltered Units

Transfers from special schools to the ordinary school system in an area with no day-schools for C.P. children were discussed by Dr. J. D. Kershaw, who outlined a project for a special sheltered unit attached to an ordinary school, so that the children in it "could go into the ordinary school and work with ordinary children in those subjects in which they could compete on equal terms, going back to the unit for any special individual teaching or sheltered activities". It was clear from comments made by various speakers that the provision of suitable facilities is a problem which has not been solved anywhere in Europe. It was also encouragingly clear that everyone was also concerned with the dangers of "institutionalisation", consequent on segregating children in residential establishments without sufficient regard for their adjustment to the real world outside the units' walls.

NEUROLOGICAL EXAMINATION OF THE NEWBORN ACCORDING TO THE WORK OF PROF. ANDRE THOMAS

PAUL E. POLANI, M.D., M.R.C.P.

THIS PAPER is based largely on the work and experience of the French neurological school of André Thomas and his collaborators, particularly Mme Saint-Anne-Dargassies. My own very limited experience is mainly in the examination of the newborn at term, and I am going to talk about this.

As a general clinical statement from the neurological viewpoint, the newborn full-term infant is a tonic animal, with a certain well-developed oropharyngeal and neurovegetative automatism. Much of the examination therefore depends on assessing tone, mainly through static postures and through direct and indirect passive mobilisation under the best possible conditions of relaxation. Static postures provide information about the balance of tone; passive mobilisation aims at assessing tone critically by reference to the three r's: resistance, range, and recoil. After tone, reflexes and reactions are the next most relevant items of neurological assessment. Reactions, for the purposes of this discussion, I shall define as complex non-obligatory reflex responses, easily modified by associated physiological states.

The newborn infant is behaviourally variable. Therefore, while the result of the neurological examination depends on pathological states, it is also greatly influenced by physiological states at the time of the examination: sleep, twilight state, wakefulness, hunger, contentment, etc. It is also influenced by the physiological events that precede and accompany birth. The *conduct* of the examination depends on the skilful assessment of

possible pathological states: to the premature, frail, sick, or injured infant some phases of the neurological examination might be damaging and should be strictly avoided.

The examination is conducted according to a segmental scheme. Most of it is done with the infant lying supine; next comes examination in the prone position; in the sitting posture; in prone and supine suspension; in the vertical position; and finally, in the suspended position with the infant grasped by the legs. I shall not describe in detail the spontaneous motor activity, or the postural behaviour, or the responses to passive mobilisation of the various segments. Suffice to say that the predominant posture is one of flexion, whether in prone, supine or vertical positions; that the head position is asymmetrical in the supine position; that there is head-lag in pulling up the infant to a sitting position; and that there is stronger extensor than flexor tone of the neck muscles.

A number of signs, reactions and reflexes are sought. The most important of these are the ocular signs, the Moro response, the grasp reflex, the cardinal-points sign, the crossed-extension response, the righting reaction, the trunk-flexion reflex, and automatic stepping. In the description that follows the normal responses will be indicated.

Segmental Scheme of Examination

Starting the examination in the supine position, the ciliary reflex is sought by lightly stroking the eyelashes—this causes homolateral or bilateral eye closure. The blinking reflex to a strong light is assessed, together with the pupillary reflex to a milder light. These reflexes are more easily observed while the infant is quiet and feeding. The nasopalpebral and the McCarthy reflexes are tested by gently tapping the root of the nose or the supraorbital region respectively: a positive response consists in bilateral or homolateral blinking. The auriculocephalic reaction is obtained by stimulating the skin of the auricle, which results in rotation of the head to the opposite side. The cardinal-points sign is sought by repeated stimulation of the angles of the mouth, the centre of the upper lip and that of the lower lip: the normal response consists in lip movements towards the stimulating finger, with tongue movements in the same direction, followed by head rotation.

The reaction to daylight is positive after the first week or so of life. The infant is confronted gently with a source of subdued light, such as that from a window. If an infant is orientated so that the light is on his right, his eyes and head will look in that direction. If, then, the infant is turned through 180° so that the light now comes from his left, his eyes and head will turn to the left. The rotation must be very slow to avoid stimulating the labyrinths. In the doll's eyes phenomenon, elicited in the first 10 days of life, the ocular bulbs stay fixed and do not follow the head when it is gently rotated to right and left and tilted to and fro along its bi-auricular axis. The jaw-jerk is normally present and is facilitated during inspiration.

Passing on to the examination of reflexes and reactions of the upper limbs, we first test the digital response. Repeated stimulation of the ulnar border of the closed hand and little fingers causes progressive extension of the digits in an ulno-radial direction, the thumb being the most refractory. On cessation of the stimulation, flexion is resumed in the reverse order. The palmar grasp and the tonic reaction of the finger flexors are stimulated by strokes and gentle pressure of the digito-palmar sulcus. The hand closes over the stimulating object, and the tonic contraction of the finger flexors is tested and its strength assessed by the ability of the examiner passively to extend the child's arm and lift, to a greater or lesser extent, the child's trunk and even its head off the couch. Normally the supinator, pronator and biceps jerks are present, but the triceps jerk is normally

absent.

The Moro reflex is constant, though it can be absent in the premature infant or when the patient is soporose. It can be elicited by various manœuvres causing extension of the head; the gentlest of these is to lift the infant by the arms, without necessarily lifting its head off the couch. Release of the limbs is followed by extension and abduction of the arms, and extension of the hands and fingers, with fanning. Often the baby cries and then the second phase follows—the embrace. This is the complete Moro response, which may, even physiologically, be to a certain extent incomplete in the newborn period, or may become progressively incomplete with advancing age.

Examination of the lower limbs reveals the plantar grasp and tonic reaction of the plantar flexors of the toes, elicited in a similar way to the palmar grasp of the hand, or even better by pressure in the first interosseous space. The crossed-extension reaction is released by stimulating the sole of the foot while the leg is maintained in extension. The other leg flexes rapidly and generally extends and adducts slowly, with fanning and dorsiflexion of the toes. I will not comment on the plantar response, except to say that while the big toe usually extends, except in the first few days, the other toes may flex; or, if they extend, they may do so without abduction. Normally, the knee-jerk is present while the ankle-jerk is usually absent. Percussion of the plantar aspect of the talus results in a quick, brief extension at the knee, followed by slower flexion.

In the suspended prone position, stimulation of the skin between the twelfth rib and the iliac crest results in lateral flexion of the trunk towards the stimulated side—a most constant reflex. The righting reaction is tested by placing the infant on a firm surface, squatting on his haunches with his feet flat against the surface. Intermittent pressure of the feet against the resting plane results in leg extension, trunk extension, and even momentary extension of the head. If now the extended infant's trunk is inclined forwards, while the baby is gently propelled and rocked from side to side to aid the disengagement of alternate legs, automatic marching will result.

The righting reaction, particularly of the trunk, can also be tested by supporting the baby with one hand, with his back against the examiner's body. The baby's body is flexed. If then his soles are stimulated by repeated dorsiflexion of the feet, the baby will gradually extend his legs, trunk and neck.

In Conclusion

We cannot yet say definitely which alterations elicited in the neurological examination, taken in isolation, have a consistently pathological significance. Nor do we know what inferences can be drawn from isolated abnormal findings, or what their prognostic value is in terms of subsequent neurological status. We are, it appears, still at the stage of finding out what is normal and what the normal variation is. On the other hand, certain changes, particularly if they are asymmetrical, persistent or multiple, have, it seems, a definitely pathological meaning. At times we meet with isolated findings so different from the average run that we feel entitled to consider them pathological. Often these changes clear up completely; they all seem to go back to normal, and thereafter the infant appears to follow a normal developmental course.

The lack of correlation between subsequent development and states considered abnormal in the early days of life is at times levelled as a criticism against the value of neurological examinations of the neonate. But do we really know whether the apparently normal older infant, who seemed neurologically abnormal earlier on, has realised all the potentialities with which he was endowed by Nature? Furthermore, and in practice more important, the prognosis given in the newborn period should never be based only on the results of and inferences drawn

from the neurological examination. Other elements—above all the natural history of the condition diagnosed—enter consideration. After all, if we see an acutely ill older child with extensive neurological signs, our confidence in his recovery will depend on the cause of his troubles. If the neurological illness followed measles we will be less confident about the outcome than if it followed mumps. So any neurological abnormality in a baby with severe jaundice of the newborn has an ominous prognostic significance, while one is less certain when a similar abnormality is seen in a baby whose mother had a long and difficult labour.

REFERENCES
Saint-Anne Dargassies, S. (1954) Etudes neo-natales 3, 101.
———— (1955) Ibid, 4, 71.
Thomas, A. and Dargassies, Saint-Anne (1952) Etudes neurologique sur le nouveau-né et le jeune nourrisson, Vol. I, Paris.
————, Chesni, Y. and Dargassies, Saint-Anne (1955) Examen neurologique du nourrisson. Paris.

NEUROLOGY OF THE NEWBORN

FIFTEEN YEARS ago Prof. ANDRE THOMAS, then 76 years of age and a most distinguished neurologist of adults, became interested in neurology of the infant. To this he has since devoted most of his time, working out techniques of examination and recording the responses of many children. In 1952, in collaboration with Mme. SAINT-ANNE-DARGASSIES, he published his *Etudes neurologiques sur le nouveau né et le jeune nourisson*, and this year, in collaboration with Mme. S. AUTGARDEN, he has written a second book, *Psycho-affectivité des premiers mois du nourisson*, recording the results of his protracted studies. These books* are not yet so well-known in this country as they will be. We are very glad that Prof. Thomas and his collaborators—YVES CHESNI and Mme. SAINT-ANNE-DARGASSIES, together with Dr. PAUL POLANI—are preparing a long illustrated article on the *Neurological Examination of the Newborn and Young Infant*, to be published in this *Bulletin*. The article will give pædiatricians and others in this country an opportunity to use the techniques Prof. THOMAS has been practising. The short paper Dr. POLANI read at Oxford is published in the present issue (p. 19) as an *aperitif* to the longer article and to encourage those who read French to study the two books.

*Both books are published by Masson, in Paris.

OUTSIZE

THIS issue of the *Cerebral Palsy Bulletin* has been grossly expanded and unavoidably delayed to allow of the inclusion of the Little Club's memorandum on nomenclature and the extensive discussion of its provisions that followed its presentation at Oxford last September. There will be one or two further big numbers containing other important papers and discussions arising from the Oxford meetings, including those on electroencephalography in cerebral palsy and on hypertonus. The intention, however, is to restrain this expansionist tendency — in other words, to keep to our original 32-40 pages—once the Oxford material has been dealt with.

DEFINITION OF CEREBRAL PALSY

THE DEFINITION of "cerebral palsy" put forward in the Little Club Memorandum and discussed at Oxford was not universally acceptable. The adjective "qualitative" was, for many, a confusing one. Sven Brandt gave this deflnition, modified from Perlstein:

> "A condition characterised by impairment of movement control due to non-neoplastic and non-degenerative pathological changes in the motor areas of the brain (which, I suppose, will include cerebellum and brain-stem) acquired before or during birth or with the first years of life."

At the end of one long meeting of a group the following definition was reached:

> A condition, characterised by a persistent impairment of control of movement and posture, affecting those parts of the brain which control them and acquired in the course of the development of the brain at an early period of life.

This was modified as follows by Dr. INGRAM and Dr. POLANI, and their new version was accepted by the group:

> A non-progressive condition affecting those parts of the brain which control movement and posture, acquired in the course of early brain development.

At the last session, Prof. B. S. PLATT put forward from the chair the following definition:

> A non-progressive disorder of the control of movement and posture appearing in the early years of life and due to disorder of the brain.

Time did not permit of discussion of this definition then.

Consideration of the four definitions quoted here show that members of the Study Group agreed in preferring the phrase "movement and posture" to the adjective "motor" used in the Little Club definition before them. They also found speaking of "early brain development" or "early years of life" preferable to mentioning any exact age. These two points are acceptable.

Consideration at greater leisure suggests that to define "cerebral palsy" as a condition of certain parts of the brain, as the first two definitions do, is undesirable. No-one would dare to *define* Sydenham's chorea as a condition of certain parts of the brain. Our knowledge of what parts of the brain control normal movement and posture is not complete and is at the moment, to some extent, in a state of flux. Nor does our present knowledge of the neuropathology of cerebral palsy allow us satisfactorily to correlate clinical observations with brain lesions. Prof. PLATT'S definition appeals because it is simple and it starts from clinical observation. But the term "non-progressive" is not suitably applied to the disorder of movement and posture in cerebral palsy because the disorder nearly always changes in the course of years. If anything is non-progressive it is the pathogical lesion, whether it is a scar or a failure of development.

We therefore suggest that in the light of discussion at Oxford and since then, the Little Club definition should be modified to read as follows:

> "Cerebral palsy" is a persistent but not unchanging disorder of movement and posture, appearing in the early years of life and due to a non-progressive disorder of the brain, the result of interference during its development.
>
> Persistence of the infantile type of motor control, such as may be seen in intellectually handicapped children, is not considered to be "cerebral palsy".

R. C. MAC KEITH
I. C. K. MACKENZIE
P. E. POLANI.

ON DISCUSSING TERMINOLOGY AND CLASSIFICATION*

MANY PEOPLE do not see any need to discuss terminology and classification in the abstract and express irritation if they are asked to do so. An illustration, such as was given at the Radcliffe Infirmary during the Oxford meetings, of the difficulties of coming to agreement on diagnosis of specific cases, and on reasons for making a diagnosis, may demonstrate the usefulness of clarifying one's ideas. But this can be a painful procedure. Disagreements and criticism tend to be taken personally, and understandably so: who likes it to seem that he doesn't know what he is talking about? Discussion, if it is of any use at all, is bound to challenge established habits of thought and, as is only to be expected, some of the criticism of, for instance, the Little Club Memorandum seemed to owe more to natural and understandable resentment to change than to detached consideration of the issues involved. The suggestion made at the end of the conference—that the memorandum should be put back in the incubator to undergo maturation—might therefore be adopted, but as much to allow further maturation to take place in the discussion's participants as in the memorandum itself.

The intrinsic difficulties of the subject were intensified not only by these emotional overtones but by failures to detect that statements of different kinds were being used as though they were of the same kind—namely, *descriptive* statements and *inferential* statements.

Descriptive statements, about signs observed.—At the Radcliffe it was clear that disagreements between different examiners about the signs observed in the cases were due to:—

(a) Disparities of meaning of terms; use of the same term for different things, or of different terms for the same things. Often a term (such as aphasia) covered a much larger number of signs for one person than for another.

(b) Difference in the situation in which the sign was elicited—e.g., stretch reflex with and without head rotated—or failure to describe the situation sufficiently.

(c) Differences of "set" or "expectation" among observers, which led them to notice, or to seek, different signs according to their (not necessarily conscious) assumptions or hypotheses.

These statements about signs observed—descriptive statements—can be validated by reference to the case itself, and disagreements resolved by re-examination. This is not so of inferential statements.

Inferential statements—i.e., statements involving extrapolations, predictions or guesses. An inferential statement cannot be checked by reference to the case itself because it involves information derived from previous experience of cases thought to be similar. It involves a guess as to the class to which the case can most usefully be assigned. There is therefore plenty of room for disagreement on inferences, both because of differences in terminology and because different people have different experiences to call on. For instance, there was disagreement as to whether or not the child shown in Dr. Epstein's film driving a tractor should be described as athetoid. One person said no,

*These notes were written for participants in the discussions at the Oxford Conference, particularly those on terminology and classification and on the cases seen at the Radcliffe Infirmary. It is hoped that they will also interest others concerned with similar problems.—Ed. Cerebral Palsy Bulletin.

because he would not expect an athetoid to be able to concentrate enough to learn to do this. In describing a child as "athetoid", therefore, he would be implying an inference as to his educability, but others would not imply the same inference.

There is a natural tendency to prefer to make inferential statements rather than descriptive ones, because they contain more information, they are more condensed. This was clear at the discussion at the Radcliffe which was supposed to be on the physical signs which participants had seen in patients whose name and age only were given, the history being deliberately withheld.

Although the Chairman often reminded us that the intention was to discuss physical signs only, little notice was taken. Several times the first statement a person made in describing a case involved an inference—e.g., "premature"; "congenital". There were frequent requests for the history of the cases—i.e., for information which might help to make an ætiological diagnosis or to validate a guess, already made, as to ætiology. Dr. McCarthy actually commented on the urge people seemed to have to make an ætiological diagnosis, but no-one took up his point and if it had any effect at all on the discussion it seemed to be in the direction of licensing the making of ætiological guesses rather than on restricting them.

It seemed then, that at the Radcliffe Infirmary, though the plan was to discuss *individuals*, there was a very strong tendency to discuss *classes*.

In both the discussions supposed to be about cases and those supposed to be about classification, the great difficulty seemed to be in agreeing on what is correlated with what.

For the clinician, the main aim of classifying a patient is to help to make a good guess about the patient's reaction to possible treatment—i.e., to get access to information about the patient which he cannot get immediately from the patient himself. The clinical examination starts from an assumption or hypothesis as to a suitable possible classification of the patient, and this determines the selection of signs that will be observed. A wider range of signs will be sought and detected if several hypotheses as to class are consciously kept in mind.

The method of examination of the patient assumes that certain signs tend to be highly correlated with some signs, less with others. To the extent that certain characteristics are known to be correlated together in other patients, inferences can be made from signs that are detected in the present patient about other of his characteristics which are not at the moment detectable (e.g., site of lesion in C.N.S.); about his past (e.g., cause or causes of the disorder), or about his future (prognosis).

An ideal classification would bring together patients who are alike in all significant respects and separate them clearly from others. Members of a class Z would have characteristics *a, b, c, d, e, f, g, h* in common. If a given patient is examined and found to have features *a, b, c, d, e*—(these characteristics being physical signs and symptoms, and perhaps features of environment, development, characteristics of family, etc.), then, if he is classified as Z, it is inferred, consciously or unconsciously, that he will also have other features, *f, g, h*, that members of the class have, but which are not at present ascertainable in this patient—e.g., morbid anatomy of the brain; or outcome of the illness, or reaction to certain methods of treatment. Classification thus enables inferences to be made. Notoriously doctors may agree on the signs observed in a patient and disagree on what inferences can justifiably be drawn.

For the research worker, the main aim of classifying patients is to help him to discover causes of differences between them (both causes of disorder and causes of improvement) by making groups homogeneous in prescribed characteristics.

As to the criteria used for classification,

it seems to be generally felt that an ætiological classification should be aimed at. An ætiological classification is felt to be satisfactory because, if a "cause" has the same effects in different people, or has some of a number of effects in different people, and has not the same effects as another "cause", then a given constellation of signs, highly correlated together, can be subsumed under the name of the cause. The great value of ætiological classification is in indicating methods of prevention (as of athetosis associated with kernicterus); it is not necessarily more useful in treatment than classification by clinical signs, though knowledge of cause may give a clue which directs further investigation and treatment (as the association of kernicterus and athetosis and speech disorder may suggest the need for examination for high-tone deafness). This is just a case of the more you know the better you can guess.

Classification by site of lesion may help in treatment (as when a diseased part is excised), but not necessarily so.

Sometimes the site, kind and extension of the lesion can be discovered in the patient; more often they are guessed at. In this case to classify by site of lesion is simply to summarise, in the name, a number of signs known to be highly correlated in many patients. When the site of the lesion can be determined, it may be that a homogeneous group is best formed according to it; but not necessarily, since in some cases lesions may be present without any morbid signs.

* *

At Oxford the English-speaking members were extremely favoured in that we did not have to translate from our mother tongue in order to communicate with our guests; but this made it the more clear how inadequate is our command of scientific language—how little we understand the implications and associations of the words we use in talking to each other and, worse still, in talking to ourselves in thought. Dr. Carmichael, in closing the discussion on classification, quoted a wise remark of one of his teachers in neurology. "Never use any words that are from another language; describe exactly what you do, exactly what you see in simple language, and there will never be any doubt what you mean." The intention of these notes is to indicate the background of some of the difficulties involved in following this advice, without discouraging anyone from trying to do so.

M. L. JOHNSON ABERCOMBIE.

THE LITTLE CLUB

MEMORANDUM ON TERMINOLOGY AND CLASSIFICATION OF "CEREBRAL PALSY"

During the past year and a half a number of interested people have met at the Little Club to discuss a memorandum on the terminology and classification of "cerebral palsy". The memorandum in its present state was put forward for discussion at the Oxford Study Group on Child Neurology and Cerebral Palsy.*
The participants in the discussions were: Dr. N. S. Alcock, Dr. J. Bates, Dr. E. A. Carmichael, Dr. J. Crosland, Dr. P. R. Evans, Dr. M. C. Joseph, Dr. M. J. McArdle, Dr. A. D. McDonald, Dr. R. C. Mac Keith, Dr. I. C. K. Mackenzie, Prof. A. A. Moncrieff, Dr. P. E. Polani and Sir Charles Symonds.
The memorandum gives a definition of "cerebral palsy", notes the need for definition of clinical terms and suggests that at the present time clinical features offer the most useful basis for classification; the clinical features of hypertonus are discussed in detail and then a clinical approach to diagnosis is put forward. This suggests that the spastic cases should first be recognised and then, from among the remaining cases, the dystonic and choreoathetoid cases, leaving a residue which will include mixed cases, ataxic cases and cases of atonic diplegia.

INTRODUCTION

In recent years "cerebral palsy" has received much attention from pædiatricians. They have made new observations about the origins and clinical course of various types. Various terms have been used and received more or less wide acceptance among them. It has become evident that not all pædiatricians have given the same meaning and use to certain terms and that there are divergencies in usage of terms between them and neurologists.

It would be helpful if neurologists, pædiatricians and others working with "cerebral palsy" would agree on its definition and on the meaning of the terms employed, for each group has something to give the others.

A commonly agreed classification of cases of "cerebral palsy" could also be helpful. Only by use of a commonly agreed and generally adopted terminology is exchange of information profitable and co-operation in research possible.

DEFINITION OF "CEREBRAL PALSY"

Because the cases included in the term "cerebral palsy" are not all identical from clinical, ætiological, and morbid anatomical aspects, the term "cerebral palsy" is not an exact one. In *scientific usage* as we learn more about each, the names of individual disease entities within the group will sometimes be preferable. Meanwhile, however, for *research purposes*, the term "cerebral palsy" is useful in the study of a group of conditions often of obscure origin affecting the brain and arising in early life. A strict definition is necessary and should be adhered to if the term is to be used for certain types of research and in the exchange of information. Finally, in *common usage* the term has administrative usefulness and a practical value largely because the affected children in many cases have certain special motor, intellectual and emotional difficulties, and so may have certain special therapeutic and educational needs in common.

*Two small changes have been made in the Memorandum: (1) The paragraphs referring to distribution of various types of "cerebral palsy" in published series has been moved to a later section. (2) In three headings the word "differentiation" has been substituted for "classification".

In the present state of our knowledge the term "cerebral palsy"[1] is usually taken to imply a persisting qualitative motor disorder due to non-progressive interference with development of the brain occurring before the growth of the central nervous system is complete.

This statement may require some elucidation.

1. Clearly only persisting and not temporary motor disorder of cerebral origin is to be included. Temporary motor disorder such as occurs during epileptic fits is not included.
2. Practically all children with a severe intellectual handicap have some motor disorder which expresses itself in a motor pattern inappropriate to their chronological age, poor motor skill at any age, shambling gait, and hypotonia. For instance, the majority of mongols show hypotonia and, for their chronological age, poor motor control. There are other groups of patients with severe intellectual subnormality that have a similar, infantile, motor pattern.

   On the other hand there are cases where to a severe intellectual subnormality is added a *qualitative* motor disorder such as spasticity, dystonia and unwanted movements which deviate completely from normality at any age. These we accept as examples of "cerebral palsy" provided the condition is not progressive. For instance, if the progress of the disorder had been arrested, a patient with phenyl pyruvic oligophrenia with qualitative motor disorder would be included.
3. Motor disorder of cerebral origin due to progressive disorder is not to be included under the term "cerebral palsy". The pathological process producing the "cerebral palsy" may act for a variable period but is limited in time. Usually it is self-limited but its cessation may be the result of treatment. The clinical evolution seen in cases of "cerebral palsy" with the passage of time has usually been accepted as being explained by nervous system growth and maturation with the resulting emergence of new functions and their interaction with old deficits. Some workers think this explanation inadequate and suggest that some of the changes in clinical findings could be ascribed to progressive pathological lesions.
4. The interference with development of the brain (the damage) may be intrinsically or extrinsically caused; before, at, or after birth.
5. The brain is taken here as that portion of the central nervous system above the foramen magnum.
6. The effect on a child of a motor handicap of cerebral origin, present from early in his development, is very different from that of a similar disorder occurring later.

For the purposes of definition a time limit has to be fixed, and in practice it is more useful to set a time limit by the appearance of a recognisable motor disturbance rather than by the time of the causative brain lesion. *The definition of "cerebral palsy" therefore proposed is:*

> *a persisting qualitative motor disorder appearing before the age of three years, due to a non-progressive intereference with development of the brain.*

DIFFERENTIATION OF DISORDERS WITHIN THE GROUP OF "CEREBRAL PALSY"

1. For administrative purposes the description of an individual case should include information regarding motor and sensory disability, speech disorder and epilepsy (if present), and intellectual and emotional impairment, while the therapeutic and educational needs should be indicated. For scientific purposes, in dealing with cases of "cerebral palsy" it is necessary to classify the disorders within this group.
2. Because knowledge of causes and

[1] It might be argued that what is considered in this memorandum should strictly be termed "infantile cerebral palsy" but as the term "cerebral palsy" has no place in the neurology of adults the adjective infantile before "cerebral palsy" is not considered necessary.

morbid anatomy is incomplete it is clear that at present classification will best be *based on clinical observations.* The main drawbacks of such a classification are that it may not correlate unequivocally with causes or with morbid anatomical findings and that, as the clinical picture evolves and changes with the growth and maturation of the nervous system of the affected child, the disorder may have to be reclassified. This defect is inherent in any classification based on clinical findings. Therefore this classification will need revision in the light of further knowledge.

3. If classification is necessary to promote useful exchange of knowledge, and if it is best based on clinical observations, it is clearly very necessary to define all terms used in clinical descriptions.

The clinical approach to the differentiation of cases of "Cerebral Palsy"

The incomplete state of knowledge of the causes and morbid anatomy of "cerebral palsy" suggests that a classification will at present best be based on clinical signs rather than on possible causes or sites of damage inferred from these signs.

To say this is not to deny that the value of eliciting neurological signs—in fact, the value of the whole neurological examination—lies in the inferences that can be drawn as to the integrity of the nervous system and its component parts; nor to deny that these, taken together with the history and checked by the results of special investigation, may allow a diagnosis to be made of the site(s) of lesion(s), and of the cause and consequently of the course, the treatment and the outcome.

We must, however, note that while it is accepted that with adult patients, because of the great body of knowledge accumulated by clinical neurologists, neuropathologists and neurophysiologists, this aim can often be reached, with children the position is somewhat different because in the field of developmental and infantile neurology comparable knowledge is scanty. Because they frequently have to be based on un-tested analogies with the neurology of adults, inferences from the findings at clinical neurological examination of infants and children are often less legitimate, less certain, than would be the case with adults.

It might also be remarked that we may be entering a period of reassessment of some of the basic ideas of neurophysiology and neurology. This is another argument for a clinical approach to classification at the present time.

If a classification based on clinical signs is to be useful, there must be a clear exposition of the bases of the classification and clear definitions of what is meant when clinical findings—e.g. spasticity, alterations in tone, etc.—are referred to.

Clinical features on which differentiation of cases might be based

In various series published to date, the distribution of the main forms (clinically classified) of "cerebral palsy" has been more or less—

Three well-recognised groups:
- cases with hemiplegia, about one-third of all cases;
- cases with bilateral spasticity, about two-fifths;
- cases with "involuntary" movements, about one-seventh;

and the remainder:
- cases with ataxia, rigidity or hypotonia, and cases difficult to classify —e.g., certain mixed forms, totalling about one-seventh.

It should be stressed, however, that some of the variations of incidence of "cerebral palsy" and the proportion of different forms of "cerebral palsy" in various reported series can be explained by the fact that individual examiners have adopted different definitions or interpretations of "cerebral palsy" and of the various clinical groups.

In order of frequency the main neurological anomalies present in patients with

"cerebral palsy" are: (1) weakness; (2) alteration in muscle tone; (3) unwanted ("involuntary") movements.

Weakness is not a useful criterion of classification because in young children, in borderline cases, in patients with dystonia, and in those with unwanted movements it is difficult to decide on its presence or absence and in the remainder it is universal.

Tone, the resistance of muscles to passive movement of the limb at a joint when voluntary control is not in action, is highly valued by neurologists for classification. Estimating abnormalities of tone requires experience. It is worth remarking that in children with neurological disorder, physical signs vary, especially as regards muscle tone,. While this may increase the difficulty of recognising abnormalities of tone it does not diminish their fundamental importance as criteria. The child should be examined under standard conditions, should be quiet, not hungry, not crying, and not asleep. For preference he should be examined in various positions—prone, supine, on his side, supported by the arms, suspended by both legs and either leg, and standing. When the child does not cooperate fully, tone can be affected by voluntary movements. The presence of unwanted movements will also affect the assessment of the tone of muscles. It is suggested that to avoid being biased the examiner should test and record muscle tone before going on to the examination of reflexes.

Posture depends, in part at least, on tone, and postural patterns, both static and mobile, should be noted as part of the examination of tone. Here again the child is to be examined under standard conditions and in various positions. Mobile postural patterns, as when the hand is about to grasp, and of arms and legs in walking and running, are to be noted as well as static postures.

Unwanted (involuntary) movements are an important criterion but are present in only a relatively small proportion of cases, and therefore are not, for the large majority of cases, useful for classifying.

Conclusion: It appears that tone is likely to provide the best basis for simple clinical differentation of "cerebral palsy". Tone therefore deserves priority in the task of definition of terms.

THE RECOGNITION AND DIFFERENTIATION OF HYPERTONUS

The biggest main group among the cases of "cerebral palsy" is that of the spastic cerebral palsies, which includes nearly all the hemiplegic cases and more than half of the remainder. The recognition of these cases, that is to say, the recognition of the cases with spastic hypertonus (spasticity), is the important first step in differentiating the different types of "cerebral palsy".

Tone is the resistance of muscle to passive movement of the limb at a joint when voluntary control is not in action. Among those cases with an increase in muscle tone we can distinguish two groups. The first, in which the hypertonus is of "spastic" character; the second, in which it shows characters of "rigidity".

1. Spacticity

In the cases of spastic hypertonus, *resistance is usually found in only one direction of movement at the joint.* Spastic hypertonus may present in various ways:

(A) Sometimes the resistance, the hypertonus, is present when passive movement is first attempted and, on prolonged steady pressure, yields slowly, the resistance being present in only one direction of passive movement at the joint.

(B) In other cases, the increased resistance, present when passive movement is first attempted, on steady pressure gives way rather suddenly and completely. (This is sometimes called a "give"). This is the so-called clasp-knife phenomenon. Occasionally such hypertonus is so severe that it cannot be passively overcome unless the position of the child is changed (for ex-

ample, by laying him on his face) or the examiner waits.

These two pictures, (A) and (B), are clearly examples of hypertonus; there is a further type of hypertonus—i.e., of resistance to passive movement—which like those described under (A) and (B) is a "spastic" hypertonus:

(C) Sometimes, when a limb is passively moved at a joint, in the course of the movement a sudden momentary resistance is felt. This type of muscle resistance to passive movement is sometimes referred to as "catch".[2] It may often be elicited in muscle groups showing the phenomenon of spastic hypertonus described in (A) and (B).

In some cases, in which spastic hypertonus has shown in certain muscle groups, the "catch" may be found in other muscle groups which do not show such hypertonus. In other cases without spastic hypertonus of the types described under (A) and (B) in any muscle groups, the "catch" is found in association with normal tone.

Distribution

Spastic hypertonus often has a characteristic distribution. In the upper limb it is greater in the adductors of the shoulder and the flexors of the elbow, wrist and fingers. In the lower limb it is greater in the adductors of the thigh, and the plantar flexors. It is usually greater in the extensors of the knee but often hypertonus can be manually elicited in the hamstrings. Posture and age make a difference in the distribution of the hypertonus. Spastic hypertonus in the extensors of the knee is found in small children suspended by the armpits but later the flexors become more involved.

Where there is spastic hypertonus as above described, there are commonly concomitant signs:

(a) clonus, which is a characteristic feature of many cases with spastic hypertonus; and

(b) increased tendon reflexes.

Clonus and/or increased reflexes which are bilaterally symmetrical can be normal and may result from nervous tension. The evaluation of the significance of clonus and increased tendon reflexes is therefore often not easy. (It is, however, probably true that the mechanism which produces the "catch" on sudden passive movement is also responsible for producing a pathological increase of tendon reflexes and clonus).

Increased tendon reflexes are most clearly abnormal when there is a difference between the limbs of the two sides. On occasions pathologically increased tendon reflexes are found in association with normal tone or with hypotonia. Pathological increase in tendon reflexes, even in the absence of spastic hypertonus, is usually taken to have the same implication as if spastic hypertonus were present.

The Plantar Response

The decision as to the presence or absence of hypertonus and whether it is spastic or not must be made on the character of the tone in the limbs. Unless hypertonus, including the "catch", as both are described above, or pathologically increased tendon reflexes, is (are) present, then spasticity cannot be said to be present. The presence or absence of an extensor plantar response can therefore have no bearing on the diagnosis of the presence or absence of spasticity or on the type of hypertonus if hypertonus is present.

2. Rigidity

In the second group of patients with hypertonus the increased resistance is generally felt in *both directions of passive movement at the joint*. This is referred to as rigidity and is of two types. In one the resistance is smooth throughout (so-called plastic or leadpipe rigidity). In the other the resistance is felt as a series of regular "catches" (so-called cogwheel rigidity).

[2]Denny Brown, D. (1946) Diseases of the Basal Ganglia and Subthalamic Nuclei. New York. p. 282.

In both spasticity and rigidity the hypertonus can vary in intensity from time to time, in subsequent examinations and in the course of one examination, especially in relation to emotional changes.

A CLASSIFICATION OF "CEREBRAL PALSY" BASED ON CLINICAL OBSERVATIONS.

Spastic "Cerebral Palsy"

Patients with spasticity form the group of *spastic "cerebral palsy"*. In this group, three main types are distinguished according to the distribution of spasticity and weakness:

(1) *The spastic hemiplegias.*—The upper limb is usually more affected than the lower. Sometimes it may appear that there is a monoplegia of one upper limb; but neurological abnormalities can be detected in the lower limb as well. At other times there is the appearance of a crural monoplegia though careful examination will reveal involvement of the ipsilateral upper limb[3]. This explains many instances of so-called cerebral monoplegia.

(2) *The spastic diplegias.*—There is affection of the muscles of all four limbs. The lower limbs are the more affected. At times the upper limbs are so little involved that the observer may at first detect no abnormality. In some classifications these last cases have been grouped under the name of "cerebral (spastic) paraplegia".[4]

Generally, however, one or more of the following signs—impairment of voluntary movement, spastic hypertonus including the presence of the "catch", and increased tendon reflexes—are to be found in the upper limb.

In cases with bilateral limb spasticity the muscles of the trunk and neck commonly appear hypotonic.[5] This *hypotonia* of the muscles of the neck and trunk does not exclude the diagnosis of *spastic* diplegia or double hemiplegia (v.i.), which is based on the condition of the muscles of the limbs.

(3) *The spastic double hemiplegias.*—All four limbs are affected, the upper limbs are more serious involved and there are usually signs and symptoms of pseudobulbar palsy.[6] The neck and trunk muscles are usually weak and hypotonic.

By some workers the spastic diplegias and spastic double hemiplegias are grouped together under the name of spastic tetraplegia (or quadriplegia); but this term is best avoided because it implies for many neurologists a high spinal origin and also because it does not reveal the distribution of the disordered function. Other workers have used the terms symmetrical and asymmetrical tetraplegia as synonyms of spastic diplegia and double hemiplegia respectively, but not all diplegias are symmetrical nor are all spastic double hemiplegias asymmetrical.

Note 1.—In a small number of diplegic patients with brisk tendon reflexes, hypotonus, not hypertonus, of the limbs is found. These cases of "atonic diplegia" do not fit in the group of the spastic cerebral palsies as above defined, but if the tendon reflexes are considered pathologically increased, the inference may be the same as if spastic hypertonus were present. In this classification the small number of severely hypotonic cases are all grouped together (vide infra).

Note 2.—Cortical sensory loss is found in a proportion of hemiplegic patients. Whether this is also true of patients with bilateral involvement, and with what frequency, is not known.

Note 3.—Athetotic movements are commonly observed in patients with

[3]For example, if fine and rapid alternating movement of hand or foot is attempted impairment of voluntary movement may be more evident.

[4]The adjective "cerebral" is necessary here because the name paraplegia has for most neurologists a strong connotation of spinal origin.

[5]Hypotonia is defined here as a decreased resistance to, and increased range of, passive movements.

[6]This term is used to denote difficulty in chewing, swallowing and talking, with weakness of the facial muscles, weakness and spasticity of the muscles of mastication, and usually an exaggerated jaw reflex. This is thought to be due to bilteral affection of the supranuclear pathways of the bulbar nuclei.

spastic hemiplegia who have reached adolescence. They are also seen in the double hemiplegias but are rarely seen in the diplegias. They appear to be a part of the picture of the spastic cerebral palsy in these cases and not to warrant labelling as "mixed" cases (v.i.).

Dystonic and Athetoid "Cerebral Palsy"

When the spastic cases have been indentified the majority of patients left show characters in common which leads many workers to group them together, usually under the name *athetoid cerebral palsy.*

This term implies a clinical picture comprising:

(1) greater or lesser abnormalities of muscle tone;
(2) greater or less abnormalities of posture;
(3) absence of spasticity, and of its concomitant signs; and
(4) usually the presence of unwanted movements (generally choreic or athetotic) in children over the age of 2 years.

For the workers who use the term "athetoid cerebral palsy" with the above meaning, [7] the concept is one of a clinical syndrome with a usual history of neonatal jaundice or neonatal asphyxia and a defined clinical natural history which evolves into the picture of a patient with abnormalities of tone and posture and unwanted movements. In early life when the unwanted movements are absent, the history, the alterations of muscle tone, the abnormalities of posture, and the absence of signs of spasticity all allow a reasonably accurate prediction of the emergence of unwanted movements at a later stage. For this reason, some pædiatricians diagnose as cases of "athetoid cerebral palsy" children who already have unwanted movements, and also those whom they expect to develop them later.

However, this is a strictly clinical classification and patients who do not show unwanted movements should not be included under the athetoid heading.

[7]Other workers have used, almost synonymously, the term "dyskinesia".

As has been said above, the majority of cases of "cerebral palsy" which do not fall within the group of spastic cerebral palsy have certain characters in common which mark them off from the remaining few (the ataxic, mixed and other rare forms). They should be subdivided into two groups. *A first group,* in which only weakness and disorders of muscle tone (and posture) are present. This group consists mainly of infants and younger children. *A second group,* in which the essential feature, usually additional to weakness and disordered muscle tone, is the presence of clear-cut unwanted movements. This group is chiefly of older patients.

In the first group tone may be increased or decreased or may fluctuate. Any increase of tone is of a plastic or cog-wheel type and signs of spasticity are lacking. Disordered static and mobile postural control is present. For the group thus defined the name *"dystonic cerebral palsy"*[8] should be used and reserved.

In the second group of patients the essential characteristic is the presence of unwanted movements. These are choreic or athetotic,[9] or frequently more difficult to define.

In some cases the unwanted movements are indistinguishable from those of rheumatic chorea, but often it is difficult to know whether choreic or athetotic is the right term to apply and the term "choreo-athetotic movements" has much to commend it. It should be noted that the shorthand description "choreo-athetotic movements" should be accompanied by a longer description of the unwanted movements in terms of their distribution, force, range, rhythm, speed and their association with various states of the patient. For instance, the unwanted movements are usually more obvious during physical or mental effort or when

[8]For neurologists in this country the term dystonia implies a disturbance of tone. It does not imply a specific clinical entity.

[9]Chorea and athetosis are non-repetitive—tremor is a regularly repetitive movement. Tremor by itself is not seen in patients belonging to this group.

the subject is under emotional strain.

Sometimes the movements show a terminal increase on approaching an object, and this increase may be regular or irregular. These patients with unwanted movements have also disorders of tone and posture, but the presence of the unwanted movements takes precedence in labelling them. Their capacity to perform voluntary movements is usually impaired. This may be due not to weakness but to disorder of tone or to unwanted movements.

The disorder of posture is apparent both at rest and in movement and it is related to alteration in muscle tone, which may be increased (hypertonus) or decreased (hypotonus) or may fluctuate between these two states. When the tone is increased, the increase is felt equally through the range of passive movement at a joint and in both directions. It is therefore rigidity, plastic or cog-wheel, and not spasticity.

At some period, during the first year of life, the muscles of the neck and spine tend to go into intermittent contraction, producing opisthotonos. After the age of one year, opisthotonos is usually no longer seen, though it may persist in children of this group who have severe intellectual impairment. With this exception, continued weakness and hypotonia of muscles of neck and trunk is usual after the age of one year, though it tends to disappear during childhood.

To describe this variety of "cerebral palsy" we feel that *choreo-athetoid cerebral palsy* is a better term than athetoid cerebral palsy and deserves acceptance.

Other Forms of "Cerebral Palsy"

Mixed Cases

Having identified the purely spastic, dystonic and choreo-athetoid forms of "cerebral palsy", a proportion of the remaining cases may be recognised as *"mixed cases"*, in which features of both spastic cerebral palsy and dystonic or choreo-athetoid cerebral palsy are present.

There are two main groups of "mixed cases": (1) those showing a mixture of spasticity and rigidity, either plastic or cog-wheel; and (2) those with spastic hypertonus who have also unwanted movements.

It should be noted that the presence of minimal rigidity in a spastic case, or of minimal spasticity in a dystonic patient, is relatively common. It would serve no useful purpose to call these cases "mixed". The predominant feature is the one that determines the clinical designation *though the accessory findings should be noted and recorded.* Similarly the presence of a relatively minor degree of spasticity in cases with marked unwanted movements (choreo-athetoid cerebral palsy) or the presence of minimal unwanted movements, in cases whose outstanding feature is spasticity (e.g. in spastic hemiplegia) does not warrant calling these cases mixed—though again *the presence of the additional clinical features should be recorded.* The label "mixed forms" is reserved for cases in which none of the diverse clinical features (spasticity, rigidity, unwanted movements) is preponderant.

Finally, other groups of cases are seen:

(1) Ataxic Cerebral Palsy

A distinction between ataxia and incoordination is not always made, but by ataxia we mean incoordination which is not to be attributed purely to dystonia, weakness or choreic movements. As we are concerned with "cerebral palsy" we naturally also exclude any ataxia due to spinal disorder. The ataxia in "ataxic cerebral palsy" is probably due either to a defect in the cerebellum or its connections or to cortical sensory impairment.[10]

In most of this group the ataxia will be due to tremor of so-called cerebellar type.[11] It may affect the arms and so interfere with their precision of movements or it may affect the legs giving rise mostly

[10] There may be cases where the ataxia is due to a disorder of cortical function but in which sensory impairment cannot be demonstrated.

[11] This tremor is characteristically only brought out by movement and is regular and usually terminal.

to a tremulous gait. It may do both. Less often, and in fact rarely, the ataxia is due to dysequilibrium so that the child has difficulty in balancing on his legs or even on his pelvis when sitting. In our experience the group of ataxic cerebral palsy thus defined is small. In some series the group is relatively larger. This would appear to us to be the result of inclusion of cases of incoordination due to dystonia, weakness or unwanted movements.

(2) "Atonic Diplegia"[12]

"Atonic diplegia" is found in three groups of patients:

(a) those with some of the concomitant signs of spasticity—e.g., increased tendon reflexes;

(b) those with some of the concomitant signs of choreo-athetoid cerebral palsy; and

(c) those who are intellectually subnormal, but do not show the concomitant signs indicated under (a) and (b).

Suggested Classification

In summary, the suggested classification is:—

Spastic cerebral palsy—
  hemiplegia
  diplegia
  double hemiplegia
Dystonic cerebral palsy
Choreo-athetoid cerebral palsy
Mixed forms of cerebral palsy
Ataxic cerebral palsy
Atonic diplegia.

The Little Club,
c/o Guy's Hospital
*London, S.E.*1.

RONALD C. MAC KEITH
IAN C. K. MACKENZIE
PAUL E. POLANI.

[12]The term "atonic diplegia" is used here because it is accepted following the work of H. Yannet and F. Horton (Pediatrics 1952, 9, 204). It is taken to imply a bilateral distribution of weakness and hypotonia.

*COMMENTS ON THE LITTLE CLUB MEMO

Based on the discussions in the Oxford Study Group and revised, as opportunity offered, by the speakers.

1. Classification of Cerebral Palsy: Yesterday and To-day

PAUL E. POLANI, M.D., M.R.C.P.
Director of Medical Research Unit, National Spastics Society.

THE PURPOSE of classification is to bring together individual units having certain determinable characters in common, with the aim of making order in the apparent chaos of individual variation. The groups so formed are subjected to study and manipulation in accordance with the object of the classification. A classification may be devised as a first step for the advancement of basic knowledge. Or it may have an essentially practical purpose as when it forms the basis for treatment. *Classification* is based on *definition*, of the individual classes and of the whole group. The ability to define presupposes the ability to lay down and show clearly limits and boundaries and "to state precisely what is comprised in and meant by . . ." (*Pocket Oxford Dictionary*). Because precision is essential, a classification must state unequivocally the means or techniques by which the classification is made and the boundaries fixed. Where there may be doubt, the techniques themselves must be clearly and precisely defined. Finally, the techniques used in classifying must be such that they can be understood and used by those whose task it is to classify.

These broad considerations and principles underly any classification and that of "cerebral palsy" is no exception. All other considerations apart it is tedious and space-consuming to have to define every term exactly, every time you speak or write about "cerebral palsy", yet at present this is frequently necessary (though unfortunately not always done) if you want people to understand what you are describing.

"Cerebral palsy" may be defined and classified with various objects; these classifications may be mutually exclusive, or mutually inclusive, or overlapping. Definition and classification may be required for teaching; or for the planning of treatment or education; or for epidemiological stocktaking; or as a background for investigation into causes. It is obvious that while for some of these purposes the definition and classification will have to be very exact, for others a certain latitude is permissible or may even be desirable. At first we may have to use wide definitions, so as to be sure that we can attract all the patients we want to study. Having done this, stricter definition and classification will have to be applied according to the objects we have in mind.

In defining "cerebral palsy", one of the most crucial points appears to be: how can we best differentiate between "cerebral palsy" with intellectual subnormality and intellectual subnormality without "cerebral palsy"? That criteria can be found for such a differentiation was shown by

*Contributed to the discussion on the Little Club's Memo at the Oxford Study Group on Child Neurology and Cerebral Palsy, September, 1958.

Hilliard and Kirman (1957), who found that about a quarter of the severely intellectually subnormal children at the Fountain Hospital also had "cerebral palsy".

Having defined a group we generally need to classify the cases within the group. For "cerebral palsy" a clinical classification appears the most practical, at the present time. But—and this appears to me the greatest need—we must set out as clearly as possible the details of the technique by which this sorting out can be done.

Freud in 1897 introduced the generic term infantile cerebral palsy. He considered it opportune to group together all the neurological conditions of children and, in the absence of pathological and ætiological knowledge, classified them symptomatically. He insisted that his was a temporary classification, born out of ignorance as to cause and morbid anatomy and devised to facilitate the process of evolution of knowledge. His clinical grouping is the one which more or less, is still followed.

Since the time of Freud's work, during the first 30 years of the century, individual clinical and ætiological entities were studied. During the last thirty years or so two new factors have focused attention on the deficiencies of our knowledge about "cerebral palsy": greater emphasis on treatment and a greater interest in the causes of these conditions. Both these factors are undoubtedly related to the general shift of the focus of attention in western medicine generally and pædiatrics in particular. With the renewed interest in these chronically handicapping neurological disorders various attempts have been made to define and classify them, partly to assess their prevalence—partly as a preliminary to statistical and ætiological studies, and partly as a clinical sorting out for treatment. One of the pioneers in this field was Phelps. In 1948 he set out a definition of "cerebral palsy". It was very widely embracing, as it included all conditions ". . . which affect the control of the voluntary motor system (through) . . lesions of various parts of the brain. . . ." His 1950 classification of the disorders within the group did little more than list the conditions. Though many workers paid lip service to Phelps' definition and classification, it is obvious from the material included in their studies that they did not follow them literally but used their own judgement, made their own reservations, and generally followed their own rules, frequently unwritten and unstated. Perlstein's definition, in 1955, was practically as wide as Phelps'.

On the whole Perlstein favoured clinical classification into three main groups—spastics, dyskinetics and ataxics—with an accessory topographical classification. Perlstein's classification has been followed by many, though his definition was not always implicitly adhered to. His clinical classification fails to indicate the techniques used in arriving at a differentiation between the clinical conditions as unequivocally as possible.

In 1955 Balf and Ingram, in defining "cerebral palsy", felt that "tacit agreement" had been reached to exclude from the group progressive diseases and transient motor disturbances. Their classification was clinical and they took into account the natural history of "cerebral palsy", particularly that of diplegia and dyskinesia.

In 1956 Minear published a paper on the classification of "cerebral palsy" based on the majority opinion of the members of the American Academy for Cerebral Palsy; only 64% of them replied to the questionnaire which had been circulated in 1953 and which serves as a basis to the statements in Minear's paper. Several definitions are reviewed and, although a conclusive definition is not given, it was decided to exclude "neoplastic brain diseases" and "the progressive neurological brain diseases". In pattern the recommended classification of individual patients is similar to that adopted for

cardiac conditions by the American Heart Association. The type of motor disturbance is recorded as well as its distribution, the cause, certain supplementary findings concerning intelligence, fits, postures, visual status, auditory status and speech ability. The desirability of recording the neuropathological finding is pointed out, though at present this is not possible. Finally, a grading of functional capacity and therapeutic needs is attempted.

One of the most serious objections to this classification is the absence of an unequivocal definition which sets the boundaries of "cerebral palsy", so that it could certainly not be used for epidemiological studies. On the clinical side there is ample room for misunderstanding and insufficient explanation of the instruments of classification.

The main point is whether such a classification can be generally and unequivocally adopted. Could a medical observer apply it and have a reasonable hope that the cases classified by him would be similarly classified by an independent observer? Could the implied definition and the classification be accepted by all workers? Has it in fact been neurologists for instance, be happy to accept it as it stands and use it?
adopted by most or even many? Would

In 1957, Kurland, who made a population survey of "cerebral palsy" at Rochester, Minnesota, found all definitions of "cerebral palsy" too imprecise for adoption in epidemiological research. As Dorn (1955) says, "statistics of ill-defined . . . ailments are not only meaningless but may also be misleading...." In his careful work Kurland thought that elasticity of definition was probably entirely responsible for the variable incidence of "cerebral palsy" reported from various countries or from various surveys in one country, yet there had been plenty of speculation about the fundamental reasons for this variation. Kurland was forced to define his own boundaries for the survey he undertook.

It is apparent, then, that the existing definitions and classifications are to a greater or lesser extent divergent, even conflicting, inadequate and wanting. Most workers are forced to make their own definitions and to classify according to individual criteria. Neurologists when referring to and describing neurological conditions in children, with one or two exceptions (Merritt 1955), do not use the comprehensive term "cerebral palsy". Even pædiatric neurologists, like Ford, shun the word.

In arriving at a definition of "cerebral palsy" and at a classification of the neurological disorders on a symptomatological basis, we, as general physicians of children, should collaborate closely with the neurologists of adults. Neurologists have a great deal to offer us. We in our turn, can contribute our understanding or stress, when necessary, the lack of understanding of causes. For instance, we can point out that the word congenital is often used when referring to causes of neurological conditions in children as if it were an explanation. I need not stress that this is not so, but is a rather obsolete way of expressing a time-relationship. We can contribute our knowledge of normal development and its deviations. But, in return, if we use neurological terms we should use them appropriately and carefully. If the meaning and definition of neurological words are not clear to us, we should consult the neurologists. My feeling is that only with their help can we reach a worthwhile and widely accepted clinical classification based on neurological signs and symptoms.

The Little Club memorandum before you represents an initial effort in this direction, the first, to my knowledge, in this country at any rate. We have received, and we are grateful, and we have given. If what we have given is not sufficient, then that is our fault. It is not a final document to be accepted or rejected as such, but, we hope, a basis for discussion, an opening gambit for contri-

bution. We can only hope that you, too, will give freely. As it stands it has been put to a brief clinical test and in our as yet limited experience we have found that it works. If I may give my personal view, it was above all useful because of the definitions of the basic instruments of diagnosis and classification. But if the memorandum is agreed upon, whether in its present or any other form, its merits or faults will only come to light by putting it to the practical test. I hope that those who accept it in principle will also try it out in the field and if necessary suggest modifications in the light of their experience.

References

Balf, C. L. and Ingram, T. T. S. (1955) "Problems in the classification of cerebral palsy in childhood." **Brit. med. J.**, ii, 163.

Dorn, H. F. (1955) "Some applications of biometry in the collection and evaluation of medical data." **J. chron. Dis.**, 1, 638.

Ford, F. R. (1952) Diseases of the Nervous System in Infancy, Childhood and Adolescence. 3rd ed. Oxford: Blackwell Scientific Publications.

Hilliard, L. T. and Kirman, B. H. (1957) Mental Deficiency. London: J. & A. Churchill.

Kurland, L. T. (1957) "Definitions of cerebral palsy and their role in epidemiologic research." **Neurology**, 7, 641.

Merritt, H. H. (1955) A Textbook of Neurology. London: H. Kimpton.

Perlstein, M. A. (1955) Infantile cerebral palsy. In Advances in Pediatrics, 7, 209. Chicago: Year Book Publishers Inc.

Phelps, W. M. (1948) "Cerebral palsy." **J. Iowa med. Soc.**, 38, 509.

——— (1950) Cerebral palsy. In Mitchell-Nelson Textbook of Pediatrics. 5th ed., pp. 1361-1366. Philadelphia: W. B. Saunders.

2. The Classification of Cerebral Palsy

PROF. RONALD S. ILLINGWORTH, M.D., F.R.C.P.

From the Department of Child Health, University of Sheffield.

THERE IS nothing to be gained by trying to alter the classification of any disease unless there is something radically wrong with the present one, and unless the new one suggested is a great deal better. I cannot see what is wrong with the classification used by the American Academy of Cerebral Palsy. With certain minor modifications, such as the occasional use of dyskinesia instead of athetosis, this is widely accepted in the United States, Scandinavia, Denmark and this country. It is the classification used by numerous workers such as Grace Woods, Skatvedt, Plum and others.

The classification consists simply of spasticity, athetosis, tremor, ataxia, rigidity, the atonic form, and mixed forms. The atonic form is exceedingly rare. I have always felt that some of the cases of the so-called atonic form are merely examples of severe mental deficiency without cerebral palsy. One can say perhaps that they are atonic because of the gross head lag in early infancy when they are pulled to the sitting position or held in ventral suspension. I have personally not seen a case of cerebral palsy remotely resembling amyotonia congenita. We have all seen innumerable cases of mentally defective children with cerebral palsy who showed gross head lag, but I do not call that atony.

As for basing the classification of cases on distribution, there is disagreement about the terms used. Quadriplegia, meaning that all four limbs are involved, is straightforward. I do not mind if anyone prefers the Greek term, and talks about tetraplegia. Paraplegia, involving the lower limbs and not the upper limbs, and hemiplegia, are acceptable, but I object to the term triplegia, because I have never seen a case. Monoplegia is exceedingly rare. I cannot help feeling that if one diagnoses monoplegia in an older child one might have thought that he was an example of hemiplegia if one had seen him earlier. I say that because very occasionally one sees an infant with hemiplegia in whom the signs almost disappear in one of the two limbs.

I never use the word diplegia, because there is a difference of opinion as to what it means. Some use it to imply quadriplegia with less involvement of the upper limbs than the lower limbs. I see no purpose myself in trying to include grades of severity in the classification. It seems to me silly to use the words "double hemiplegia", when quadriplegia says the same thing more briefly.

Any classification of a disease must be simple and intelligible to the majority of people using it. Some of the classifications suggested are to my mind quite unintelligible. Their use would merely lead to wide differences of opinion and interpretation. I have not seen any classification better than that of the American Academy of Cerebral Palsy and therefore I see no purpose in trying to introduce a different one. If we do, no-one will take any notice.

---

3. Our Point of View on the Standardisation of Terminology

PROF. DR. ADELAIDE GRISONI-COLLI
From the Neurological Institute, Milan.

THIS SUBJECT is definitely less important than other research in the clinical field. But seemingly minor issues often have their place. A standardisation of terminology seems to be necessary in this field, which is growing in prominence, interest, and practical applications. It should be done by specialists of different background and from different disciplines, neurologists, pædiatricians and orthopædic surgeons working together.

Today, we find that each patient requires something more than a simple clinical classification and each presents individual characteristics that become very important for evaluation and treatment. In particular, a revision of the terminology applied to the infant patient is really indispensable, as he presents a clinical picture that differs from that of the older child on which today's classifications are based (spastics, athetoids, ataxias, mixed cases). We know, for example, that the phenomenon of athetosis does not manifest itself in cerebral palsy before a certain age (from 1½ to 2 years) but the hypotonia present in the infant with future athetosis is different from, say, the hypotonia which is followed by ataxia. In the first patient the major synergies seem to be more involved, while in the second the hypotonia seems to be more evenly distributed. We are trying to define more precisely these important details, but we meet a certain difficulty in the standardisation of the clinical picture when classifying it. It is certain that, when confronted with the infant patient or the so-called "mixed" cases (also in the second infancy between 3 and 6 years), we are often unsure how to summarise clinical pictures into convenient schedules. We are also uncertain how to classify many cases in those syndromes that are now undoubtedly undergoing revision, such as Foerster's syndrome of "atonic-ataxia", which seems more a temporary clinical phase than a definite and stable clinical entity. For these reasons the problem is a challenging one. The question is, how to handle it? The first thing would seem not a change of classification but rather a detailed definition of signs and symptoms with more careful terminology. To make the class-

ification more precise a better arranged terminology with a glossary would be of great assistance.

Undoubtedly the first common inexactness is in the use of the word spasticity, with which we label a large number of our patients who could be more exactly defined as "dystonic". In fact, apart from the cases of hemiplegia (in which spasticity often, but not always, manifests itself in the classical form), the majority of our spastic reveal, as a common characteristic, a disorder of motility characterised by hypertonia with irregular and inconstant distribution of tone both quantitatively and qualitatively. We can no longer call this hypertonia simple spasticity because it belongs to the complex and wider chapter of the alterations in the regulation of static and dynamic tone, which regulation is the function of the extrapyramidal system in which we must specify the fractions. However, in these cases, more than the spasticity, the dystonia demands our maximum attention because its type and distribution characterize each case. Besides, in the infant (until 1 year or a little over) the hypotonic component is usually dominant, especially in the zone of the "axe corporel", and dystonia of the limbs may be present in flexion or extension or torsion. These are our spastics, when babies. The same cases change under our eyes. The pathological distribution of tone can assume a more torsion-like character (especially in the upper limbs), or more similar to attitudinal dystonia, well evident in comparing the results of examination with the patient lying and standing. It can also take on the characteristics of athetoid dystonia, as can be seen in some hemiplegics who are so from earliest life. This change of the external manifestations of neurological phenomena is logical in cases where the clinical picture evolves; not because the illness is progressive, but because of the progressive maturation and functional activity of brain levels. More often we find signs of a complex functional disorder that involves different nervous structures and, in addition to the signs of self-functional impairment, reveals the phenomenon of diaschisis. We must consider these various points in modernising our terminology, so that we may have a better understanding of these difficult steps.

In other cases a disquieting characteristic is the mixing of different symptoms and signs of pyramidal and extrapyramidal impairment in different ways in each case. We can, however, reach a clearer understanding of this point since the intense studies of neuro-physiologists have afforded us a more definite explanation. I do not think that classifying these cases as "mixed" satisfies us today. For these reasons, it would seem useful to focus on the problem which, apart from the theoretical interest, presents a practical interest. This is in the application of these studies to rehabilitation, with the aim of adopting better treatment techniques for the continued improvement of the patient.

In clinical classifications, as they are at present, pictures are derived from semeiology. We talk of spastic, athetotic, and ataxic types, rigid and mixed cases, but we have seen that spastic types (apart from a group of hemiplegias) are more complex. We know that in the group of athetosis, apart from a certain proportion of pure cases, we find cases that are extremely complex. The presence of athetosis unites them, but these patients sometimes have a dystonia or impairment of tone that I do not think is any less important than the athetosis, particularly because of the value and meaning of this phenomenon in choosing methods of rehabilitation. We must not forget that a detailed study of the meaning of certain irregularities of tone may really enable us to provide rehabilitation for a case of athetosis with dystonia by using a method differing from that employed in the rehabilitation of pure athetosis.

Also, in the ataxic group, we often find cases of incoordination with or without tremor, and these cases are undoubtedly

different from each other. We have again cases of ataxia with hyptonia and other cases with hypertonia especially in the anti-gravity system. Even excluding evolving cases, which are very complex, the group is extremely varied.

Is it convenient or not to complicate the matter? That is, shall we insist on maximum precision of detail in diagnosis? For us, diagnosis means constructing a rehabilitation programme with different methods depending on the interpretation of the various neurological features of the individual patient. This is of incomparable importance to the patient. Therefore, a little detail may be more important than a general definition. For this reason it seems only just to tackle trials and tribulations in order to improve our existing classifications. A precisely detailed terminology would be invaluable for noting changes in the clinical picture during the long process of rehabilitation. As in musical composition, where a tonal change allows the use of the same note with a varied harmonic result, so perhaps in this field, with the detailed study of muscular tone in a static and dynamic context, we can give the key for a terminology that keeps pace with the changing clinical picture of each case.

In this research, the meaning or inference drawn from the pathological phenomenon is more important than a simple description of the phenomenon itself, and the study and definition of each abnormal finding should not refer to the semeiology of adults but rather to the stages of motor organisation from birth on. For example, only from the detailed description of the motor organisation can we classify the cerebral palsied infant who cannot be named as spastic, athetotic or ataxic because this terminology does not adequately define the peculiarities of each case. There are neurological signs that cannot be seen at an early age and are evident only later. But we must find a way to describe the existing condition that would give us a complete view of the case according to the laws of neurophysiology. In a certain sense, we must construct a "growing motor scale" to use as a test. Each case fits into a determined position on the scale, allowing an acceptable standardisation in a way that describes the individual case in his unique clinical picture, even when the picture changes over a period of time or during rehabilitation. Much as we evaluate a feeble-minded patient, adding to the diagnosis a more detailed evaluation by using the I.Q., so in our cases we should add a "level of motor organisation" that would more precisely define each case. These details would be invaluable in making a rehabilitation plan.

We are here in a field that differs completely from that in which we use motor tests alone.

In practice, it is important to observe not a crude expression of abilities, but rather the stage of organisation (without evaluating the more or less normal result of motor growth). In our cases the abnormal responses are interesting and significant. They are signs of functional activity, even if it is more or less disturbed or diminished. The responses always indicate levels or organisation and types of impairment.

VARIATION OF TONE

In view of the foregoing, we may summarise the impairment of muscular tone. beginning at the lowest level:

1. Association with posture, as in fixed dystonia, with greater or lesser hypertonus, which can be generalized, as in decerebrate rigidity.
2. During postural attitudes, as in static reflexes and reactions and statokinetic reflexes and reactions, both segmental and global.
3. In the course of synergies.
4. During automatisms (static and locomotor).
5. During the performance of voluntary movements.
6. During the performance of functional acquired movements (as in exercising and learning).

The difference between tone associated with posture—static tone—and that found in postural attitudes is as follows: In the first the distribution of tone is that found at absolute rest; in the second this basic static state of tone is altered by the release of static and statokinetic reflexes.

Depending on the level of organisation, the characterisation of the individual case is towards different forms that imply different physio-pathological mechanisms and have different diagnostic and prognostic meanings. The lower the scale of organisation the more severe we can call the case. We know in fact that cases which present tonic postural dystonia are less favourable in prognosis than those showing dystonia in postural attitudes. We also know that some automatic motor activities (walking, for example) necessitate a good level of organisation of global synergies. Therefore when we plan rehabilitation, we build up a progressive pattern of movement, from the simplest to the most complex, according to the summary indicated above. The different methods are adapted to the needs of the case, examined in the same schematic order. Of the different techniques available that one is chosen which is most useful in overcoming the impairment of motor organisation at a determined level.

We are not yet able to make a completely graded system of examination that embraces all the many components of reflex, automatic and voluntary movements which should be taken into consideration.

This is only a suggestion; our point of view. With this we plan to conduct systematic research to find a standardisation of terminology that would permit us to define each case in neurological language in such a detailed and graded way as to make it easy to follow his evolution step by step.

---

4. Some Remarks on Terminology and Classification

SVEN BRANDT
Director of Cerebral Palsy Clinic, Orthopedic Hospital, and of E.E.G. Laboratory, Queen Louisa's Hospital, Copenhagen.

WHEN I received a copy of Professor Illingworth's death-sentence to the memorandum a few days ago I got scared and said to myself: What am I to do? Try to act as a life-saver? Or play the part of a clergyman assisting at a burial service?

Unfortunately I know nothing about the pre-history of the memorandum. For some reason its many celebrated collaborators must have been unsatisfied with most earlier suggestions on terminology and classification. I have been surprised not to find any reference in the memorandum to much of the work on classification done in other parts of the world†. Professor Illingworth has mentioned the work done by the American Academy of Cerebral Palsy, published by Minear, chairman of the Nomenclature and Classification Committee, A.A.C.P. (*Pædiatrics*, 1956, 18, 841). And in Scandinavia classification was discussed at the First Scandinavian Conference of Cerebral Palsy in Oslo in 1954

†A 48-page memorandum, **Some Landmarks in the Study of "Cerebral Palsy" and Involuntary Movements; Definitions and Classifications of "Cerebral Palsy"**, prepared by Dr. Paul E. Polani for the Little Club, was circulated to all participants at Oxford and will be summarised in this Bulletin later.—Ed. C.P. Bulletin.

but this was in a strange language, inaccessible to the authors of the present memorandum (*Foren. f. spastisk lammede børn, Copenhagen*, 1954, *p.* 17).

I agree, however, that classification is rather difficult and that my own classification changes with time. This is a result of growing diagnostic experience and the discouraging discovery that many cases of C.P. still fall more or less outside the classes. Therefore I cannot fully agree with Professor Illingworth that time is wasted on discussions on terminology. I hardly feel that the memo's suggested classification solves the problems that confront those who struggle to use the classification given by modern pioneers in cerebral palsy. Personally I use my own diagnostic skeleton, which aims at supplying anyone interested with a quick survey of the following feature of each case: cause; distribution and type of incoordination; tentative localisation of lesion; severity; and complicating symptoms and signs. I think this is in fairly close agreement with the A.A.C.P. classification.

Should we really aim at a common terminology and classification in all details? One purpose of breaking up the general diagnosis of "cerebral palsy" into more specific classes is the hope, through various correlations with either ætiological factors or therapeutic results, of arriving at conclusions which may be important in prophylaxis or which may provide indications for special treatment. Would not such correlations possibly be obscured if we all felt obliged to stick to the same classification, at the present stage of the still recent history of cerebral palsy?

The definition now offered by the Little Club will, I fear, lead many people, both inside and outside the medical profession, to ask for a supplementary definition of "qualitative". I should like to claim that the hypotonia shown in mongols is "qualitative" too—very distinct and different from the "non-qualitative" motor retardation found in many other mentally retarded infants. So, unless you explain to us what this special quality must be, the definition given will include mongols, and hundreds of parents will then apply to your clinics for treatment.

I agree that neoplastic and degenerative disorders should be excluded by the definition, and that the term "non-progressive" in the very great majority will cover the aim, but I nevertheless feel tempted to ask whether or not some progression caused by very slow glial proliferation—as in some epileptics—may occur in a few cases. I, at least, have seen some children severely handicapped by cerebral palsy where some progression could hardly be explained in any other way.

On the other hand, I would hesitate to use the term "persistent" disorder in a definition of cerebral palsy. In my clinic an increasing amount of very mild cases of cerebral palsy are diagnosed with mild retardation of motor behaviour. To say that such cases of definite but mild cerebral palsy are *persistent* is incorrect. This is irrespective of the fact that the brain *lesion* causing the symptoms in early childhood will naturally persist.

As a definition, perhaps rather more general but explaining my own views about the condition, I prefer the one given by Perlstein (*Nervous Child*, 1949, 8, 128), but somewhat modified by my own needs and suggestions:

> "A condition, characterized by impairment of movement control, due to non-neoplastic and non-degenerative pathological changes in the motor control areas of the brain" (which word, I suppose, will include cerebellum and brain-stem), "acquired before or during birth or within the first years of life."

I prefer this last rather loosely defined upper time limit to the more categorical "three years", suggested by the Little Club. Otherwise some day the sponsors of your clinic may hang you up if you start treating a child who happened to drop out of the window on his third birthday and had a spastic hemiplegia as a

result of his fall.

The Little Club rightly emphasizes the clinical approach to a classification as the most convenient in the present state of knowledge. I agree that neither cause nor site of lesion provides a practical alternative.

Tone has been preferred as the fundamental basis for classification. I should prefer to make one more step and suggest "*incoordination*" as a common factor for all classes of cerebral palsy.

"Palsy" means weakness. But what is actually paralysed? Muscles? No! Movements? No! What has been thrown out of normal function and therefore paralysed is *movement-control*, which is what we mean by *coordination*. This incoordination may be spastic, athetoid, ataxic or "dystonic", or it may be hypotonic. Rigidity is also a type of incoordination although its identity is often confusing to me. Actually it seems to me, that neither choreoathetosis nor ataxia are placed in their right order in the outline of classification given in the memorandum on page 35. The former ought to be placed in a group called "normo-tonic"—perhaps hypotonic—and the latter should by all means appear in a group called "hypotonic". So far as I can see, the conclusion on p. 30 ("it appears that tone is . . . the best basis for simple clinical differentiation . . . ") has not been followed to its logical conclusions. What is common to the groups suggested is some type of abberation from normal coordination.

I have been rather disappointed to find no comments in the memorandum on the type of hypertonia usually called "rigidity", except for a few lines on p. 31 giving us the impression that there is no problem. I honestly admit that this is my greatest diagnostic headache, and I should like to try to clarify some of these troubles.

It seems to me that the term "rigidity" has been simplified by the Little Club, its only difference from spasticity being that here hypertonus is present in muscles on both sides of the joint. Consequently the term "rigidity" has completely disappeared from the classification. You will not find the word on p. 35.

This is certainly an easy way to get rid of an irritating problem. The trouble is that "rigidity" is a term used in various ways: the *pædiatrician* and his ward nurse talk about "rigidity" when a baby behaves more stiffly and is less relaxed than average during handling, feeding, washing and clinical examination. The *neurologist* has a definite idea of something quite special when he talks about rigidity—namely, the hypertonus found in patients with Parkinsonism, either combined with paralysis agitans or as part of the picture caused by chronic encephalitis. In children with cerebral palsy this type of hypertonus is exceedingly rare. It covers the picture described by Ford in his textbook as "juvenile paralysis agitans of Hunt" (3rd ed., p. 394). "Lead-pipe" refers to the resistance against passive movements found in this rigidity and in no other type of hypertonus. I do not feel that the "catch" phenomenon would be detected in such patients. To me, "catch" is the central characteristic of spasticity. However, it should be admitted, in support of the memorandum, that our neurophysiologists have so far been unable to verify any fundamental difference between the mechanisms of rigidity and of spasticity, except for the lack of electromyographic silence in cases of rigidity when passive movement ceases.

To some *neurophysiologists* the term "rigidity" actually means no more than increased tonus. We find the word in the condition called "decerebrate rigidity". But this rigidity is certainly of another quality than the lead-pipe rigidity. It is variable, influenced by positional reflexes, and perhaps even caused by *increased* postural reflexes.

It is tempting to suggest, as something beyond doubt, that this last condition *must* be present in a more or less complete form in many of our severely affected children with cerebral palsy. Probably the terms "dystonic cerebral palsy" or

"mixed forms of cerebral palsy" will be chosen for such cases by members of the Little Club. Eirene Collis and her collaborators would use the term "variable rigidity", which is—as far as I can see—the best name so far.

I feel unhappy about the memo's use of "dystonic" cerebral palsy in this sense. Actually this word means no more than "abnormal tone". Spasticity is equally a type of dystonia. But, under the influence of the neurological picture called "progressive muscular dystonia", this word has acquired, at least for me, a certain specific meaning, even when found in patients not suffering from this progressive disorder. I use the term "dystonia" when I find during the clinical examination a type of severe active and enduring contraction released by any attempt to perform passive movements of the joint controlled by the affected muscles. This contraction will further increase whenever the patient tries voluntarily to relax his muscles. It is no myotonic phenomenon. It is a disturbance in the voluntary control of release. You have the feeling that the voluntary intention is signalled to some postural integrative system. The energy has to flow somewhere, and since the normal riverbed is not available, it flows along paths intended for other purposes. This dysfunction has a subcortical origin, the lesion probably lying in the diencephalic motor nuclei. Most cases with this rare dystonia are hemiplegic, but there may be some cases, called "tension athetoids", who have apparently normal cortical function but are tetraplegic cases of this dysfunction.

In "variable rigidity" (which probably covers dystonic cerebral palsy in the sense suggested by the L.C.) the total involvement seems even more complex. Cortical mechanisms seem not to be intact. Many of these children grow up to disappoint the premature impression of fairly intact intellectual functions. If you have a dystonia caused by subcortical anatomical changes in such cases, this is only part of the complex decerebrating lesion.

I have mentioned cases of dystonic hemiplegia. Somebody may have seen a case of hemiballismus, and I should be surprised if nobody has, occasionally, diagnosed a case of hemiataxia.

Therefore I will have to react against the suggestion of the Little Club to limit the use of a topographic diagnosis to cases with true spasticity. I also find it very arbitrary to neglect the term tetraplegia and even to replace it by "double hemiplegia". Does a blind person have a "double hemianopia"? And why not replace the diagnosis of "gigantism" by "double hemihypertrophy"? And we at this conference, who are halfway pediatricians and half-way neurologists, should we not accept the time-saving title of "double hemi-specialists"?

Many cases of generalised spasticity do not show more severe involvement of the upper than the lower extremities. Such cases would be called "diplegias" and not "double hemiplegias" by members of the L.C., but this is again a break from earlier terminology, which many have found highly practicable. Whether "tetraplegia" or "diplegia" should be preferred may sometimes cause some hesitation. I always prefer diplegia for cases with involvement of four limbs, but only very mildly affecting the arms. This is a practical terminology for the educators and the rehabilitation experts who have to care for the child's social future. A person with a diplegia can use his arms for many purposes and may even benefit from some vocational training. The tetraplegic youngster with a moderate or severe involvement of the upper extremities is much more incapacitated.

In my opinion the smile-releasing term, "double hemiplegia", should be reserved for those rather infrequent cases where both arms are definitely more severely involved than the legs, which are only mildly affected neurologically.

I have never before realised that some workers identify *symmetrical* tetraplegia

with spastic diplegias and *asymmetrical* tetraplegias with "double hemiplegias". This is new to me. (See p. 33 in the memorandum.) But for my own part, when trying to correlate causes with symptoms, I have found it useful to classify bilateral cases into symmetrical (when both sides seem equally affected) and asymmetrical (when the neurological findings are more pronounced on one side). In 99 cases of symmetrical spastic diplegias (and paraplegias) I found the highest correlation with prematurity (50%) and the lowest correlation with neonatal asphyxia (12%), as compared with similar data (27% and 23% respectively) for all types of cerebral palsy (628 cases together). This difference was less pronounced in a group of 64 asymmetrical spastic diplegias (44% premature, 20% asphyxiated) (*Danish med. Bull.* 1958, *p.* 47).

In Conclusion:

I am inclined to think that many will feel more confused about several questions of terminology and classification after reading this memorandum than they were before. I have wondered how this might happen when so many prominent people tried to coordinate thinking. The reason seems to be found in the situation that all of us lack in what might be called "common experience". We have grown up with very different basic educations. Those of us who are pædiatricians sometimes lack the background of general neurological terminology. The pædiatricians also sometimes forget that children must have a life beyond childhood, and that diagnosis and classification might well indicate to those caring for rehabilitation how serious and how widely distributed the handicap may be.

My words may be taken as a reproach to the pædiatrician. This I would regret. How much the memorandum leaves to the influence of neurologists, orthopædic surgeons and physical medicine experts I do not know. But one thing is obvious to everybody. The problem has been opened by pædiatricians. It is this profession that has felt the need for supplementary knowledge. I cannot agree with Professor Illingworth's implied suggestion that the memorandum has been put forward as a substitute for accepted terminology and classification. To me, it has been the expression of a great need for an exchange of opinions among people who all want to see clarified some of those many deviations from textbook descriptions which they meet in their daily work with "cerebral palsy" children. This memorandum is a new-born baby, brought into the world after a difficult labour, and—perhaps—somewhat premature. It should be placed in an incubator. Some day it will grow up, and Professor Illingworth will feel very happy to see that the child acquires an increasing likeness to his Uncle Sam overseas.

5. The Use of Neurological Terms in Cerebral Palsy Studies

IAN C. K. MACKENZIE, M.D., F.R.C.P.

Physician in Department of Neurology, Guy's Hospital, London.

I HAVE been asked to follow Dr. Brandt with a few explanatory comments, some of which will be general and some in reply to points raised by him.

My own experience of cerebral palsy is relatively small and largely second-hand, in that I have never made (or even missed) the diagnosis, and have been responsible for advice regarding the further management of only a few patients in whom a diagnosis had already been made. I am therefore in no position to disagree with any classification, but some of the collaborators and myself were led to be-

lieve that an attempt to discuss a new one might have beneficial results, and our own particular function was to attend as clinical neurologists, who were ordinarily largely dealing with adults and if with children, not with those with cerebral palsy. It was clear that cerebral palsy was a neurological condition, the literature of which was naturally teeming with neurological terms, many of which had a place in adult neurology, although cerebral palsy, at any rate as a diagnostic problem, had not.

With regard to the question of classification, there is no purpose in classifying merely for its own sake, as one might collect stamps and classify them in an album. Mr. Pollock said yesterday that there are only two purposes in trying to produce a commonly accepted classification—to be able to communicate sensibly with our colleagues, both at home and abroad; and to ensure the best possible treatment for the patient.

This, in itself, seems to me reason enough to strive to such an end; but surely a further and even greater advantage might accrue, and this is that we might thereby hasten the understanding of the basic nature of cerebral palsy and its various causes. There are surely many of these—that due to Rh-incompatability is but a recent discovery—and the various causes may well give rise to different clinical pictures.

The absence, at the moment, of specific treatments is no reason why we should not, at this stage, try to become familiar with the details of such clinical pictures, which present under the broad heading of cerebral palsy.

May I quote, as an illustration of what I have in mind, that the term "paraplegia" was in common use for a variety of complaints in adults in the middle of the last century? And it may interest you to know that Addison, at Guy's Hospital, gave one lecture a year on the subject. Later different types were recognised, including a spastic type, and then an ataxic type, and eventually, at the turn of the century, by the use of a strict neurological discipline, subacute combined degeneration of the cord was identified among the paraplegias and described as an entity. The early description was certainly incomplete but the seedling flourished, so that a quarter of a century later, when the specific treatment for this condition was discovered, the clinical picture was fully recognized and it was at least known to what clinical syndrome this treatment could be hopefully applied.

Turning again to the classification of cerebral palsy, I am informed, as you have been, that the classification of the American Academy for Cerebral Palsy is not so universally accepted as is supposed, and one of its main defects is the absence of any clear definition of the terms used.

Take, for example, spasticity. If I may quote from the special article by Minear, under the heading "types of cerebral palsy defined" one finds:

> "A. Spasticity, characterised by a lower threshold of the stretch reflex. There is a large reflexogenic area, augmented responses with clonus and an abnormal electromyographic record. The pathological stretch reflex must be present to make the diagnosis of spasticity".

Now the stretch reflex is, as you all know, a physiological concept and is concerned with very complex problem of muscle tone. So seeking elsewhere, because it was not in the article, to find some description of how the pædiatrician might detect its presence and determine its departure from normal, I turned up Frank R. Ford (1952) and was delighted to find stretch reflex in the index. I turned to the page indicated and found myself in a section headed "Irrigation of the Bladder". Reading on undaunted, I came to, and I quote:

> "Lesions in both para-central lobules, such as may occur when the superior longitudinal sinus is occluded, in both internal capsules as in bilateral hemiplegia, or in high transection of the

brainstem in experimental animals, cause similar but much more severe disturbances. During the state of shock, the bladder is distended and there is overflow incontinence. Later the bladder becomes small and empties itself as soon as it is partially distended. The stretch reflex is increased".

Thinking I was perhaps on too broad a front, and rather out of date in 1952, I turned to Dr. Grace Woods' book, published last year. Alas, the stretch reflex was not indexed. In the text, however, it was said that usually the reflexes were exaggerated in spasticity owing to the stretch reflex, but this simple comment was preceded by the view that the normal examination of tendon reflexes was not very helpful. With this we incidentally agree, but we feel that careful clinical examination, difficult though it may be, could be helpful now, or retrospectively in the future, and it is within the framework of such an examination that we have discussed the words that might be used in clinical descriptions, and their meanings.

Words sometimes change their meanings through the years and sometimes have changed meanings thrust upon them. This seems to me to be the cause of one of the main difficulties in discussing cerebral palsy at the moment, and I would like to refer briefly to how such confusion may arise and how I hope it can be averted now in a particular instance.

Epilepsy is a clinical manifestation and our understanding of the epileptic phenomena is largely based on, and has developed from, the observations and writings of Hughlings Jackson. By a stroke of imaginative genius he postulated that the patho-physiological counterpart of an epileptic attack was a sudden occasional excessive discharge in the grey matter. Some fifty years later, such sudden, occasional excessive discharges were indeed seen in the EEG, but they are surely not epileptic discharges, as we have heard them called. Even if there was 100% correlation between the EEG discharge and the attack it would be wrong to use the term; and to say that a certain percentage of children had epileptic discharges in the EEG with no clinical epileptic seizures is simply bedevilling the meaning of words.

It is as if the machine is expected, almost overnight, to supplant the accumulation, through the years, of clinical experience, patiently and thoroughly collected; and it is as if you were saying to a child "my machine says you are epileptic; please have the goodness to have a fit so that I can get my figures straight". I am not for a moment detracting from the importance and the interest of the observation that some patients have sudden excessive discharges in the EEG and have so far had no epilepsy, but we must not too hastily jump to conclusions which can so easily lead to confusion, not only in terminology but in our understanding of the mechanisms involved.

I would also utter a word of warning against putting the E.M.G. on a similar pedestal rather than regarding its findings as a possible humble contribution to the sum of clinical and other experience.

I cannot review all the interesting and helpful comments that Dr. Brandt has made, because I have no experience to do so. He has, for example, touched on the "qualitative" nature of the motor disorder, and perhaps further discussion of that will come if it is necessary. With regard to his comments on tone, its abnormalities are really useful in adult neurology only if they are in the direction of hypertonia. Hypotonia certainly exists but is almost inevitably accompanied by other clinical signs which overshadow it, so that it becomes an accompanying rather than a leading sign. One may, for example, find hypotonia early in the clinical examination and then find that the diagnosis is eventually one of myopathy or of tabes or a cerebellar lesion, which makes the hypotonia as such an interesting but diagnostically unimportant observation.

There is no such thing in neurology,

so far as I know, as a hypotonic adult patient being a diagnostic problem.

The same is not true of spasticity, or of rigidity, which have important localising features—the main value, as you know, of physical signs being to localise lesions. I do not think the term rigidity has been "simplified", as Dr. Brandt suggests, but its important difference from spasticity is not only that it is present on both sides of the joint but that it acts throughout the full range of movement; and if there is not much rigidity in the memorandum there does not seem to me to be, at any rate in my small experience—and Dr. Brandt agrees—much rigidity of this nature in cerebral palsy. We should, however, perhaps have said that rigidity is constant in degree and without "give" or "catch".

I am not happy about the term "variable rigidity". This is not a prominent feature in adult neurology apart from the obvious influence on tone which may be expected, for example by emotion, manipulation and other things, and there is here reason to believe that the variability depends partly on emotion and partly on superimposed voluntary albeit incoordinate movement. Any underlying disorder of tone certainly does not seem to be rigidity as we have described it in the memorandum.

Dystonia seems to mean many different things to many different people, and it was a revelation to me to hear someone say the other day "It doesn't matter whether the tone is normal or abnormal, this is dystonia", the diagnosis apparently being made visually. Dystonia means, as Dr. Brandt points out, simply a disturbance of tone, and while there is no reason why it should not have more specific meanings to different people, they must surely be related fundamentally to some disturbance of tone, and disturbances of tone cannot be seen, they can only be felt.

Dr. Brandt finds it arbitrary to neglect the term tetraplegia and replace it with double hemiplegia. He asks: "Does a blind person have a double hemianopia?" The answer is that, if his blindness is cortical, he does, and this term has the same sort of localising value as we intend the term double hemiplegia to have. In view of the fact that tetraplegia strongly suggests to neurologists a localisation in the cervical cord, it seemed to us better to avoid its use in describing palsy of cerebral origin.

Dr. Brandt finally spares the smile-retaining term so long as it is used in its own arbitrary way. Well, why not? It is agreement we are looking for and we have no wish to impose any narrow view on anyone.

At the clinical meeting I saw a lot of tests performed which I could not either perform or interpret myself, but no consistent use made of the ordinary routine neurological examination, even so far as it could be applied, and this, I think, is a pity.

May I, in this connection, return to what I said above? Making diagnosis visually is putting neurology back 150 years. When Parkinson described the "shaking palsy" he described only what he saw. Not till 50 years later did Charcot comment on the rigidity we accept as an essential part of the typical clinical picture. The neurologist's diagnosis, whether of spasticity, dystonia, parkinsonism, or cerebellar disorder, is not made without an ordinary complete routine neurological examination. To omit this in dealing with cases of cerebral palsy seems to me, as I have said, a pity.

Mr. Pollock has since said that he thought that there was 90% agreement in the subsequent discussion. If this was indeed so, then as far as I am concerned people were speaking in code. I believe this is really what was happening. There is not nearly so much disagreement as there seems to be, and as long as it is understood that one uses dyskinesia—another word whose meaning is also bedevilled—where another uses dystonia, and one uses dystonia where another uses

athetoid, the problem of terminology will gradually move towards a solution.

I think the memorandum did not make clear that we were really much more concerned with terminology than with classification. It is not a final document but an attempt to resolve some of the difficulties mentioned. We are only at the beginning of what one hopes will soon be a most exciting further advance towards solving some of the outstanding clinical problems of cerebral palsy. "United we conquer, divided we fall" should be the watchword.

I myself feel privileged to have been asked as an outsider to make these comments, which you must accept as the sincere, if not necessarily solemn, beliefs of a deeply interested collaborator.

WORKING-PARTY DISCUSSION

Report by DR. T. T. S. INGRAM (Edinburgh)

THIS DISCUSSION opened with a review of the confusion that existed in the terminology of cerebral palsy today. It was suggested that the present use of terms was unsatisfactory, not so much because of a different choice of terms by different clinicians, but because of the different things that were meant by the same terms. There seemed to be a greater variation in the meaning of terms used as nouns than those used as adjectives. For example, the term "athetosis" tended to have a wider connotation than the term "athetoid movement". Some of the reasons for the present variations in the scope of terms in different groups were discussed. The following reasons were suggested.

Firstly, most of the terms in common use in cerebral palsy had been borrowed more or less directly from current neurological terminology designed to describe the conditions affecting the nervous system in the adult. Dr. Ritchie Russell pointed out that many of the manifestations of cerebral palsy in the child occurred very rarely, if at all, in adult neurological practice and in this sense the direct borrowing of terms was unlikely to result in accurate description. *Secondly*, the terms had been borrowed by clinicians and therapists who were interested in cerebral palsy from very different points of view. For example, physiotherapists and orthopædic surgeons would be more interested in terms from the point of view of deciding whether it was amenable to treatment or not, whereas neurologists would probably be more interested in it because of its effect on brain functioning; thus terms such as spasticity and rigidity might mean very different things to these workers. *Thirdly*, whereas the terms borrowed from neurology were in general designed to be descriptive, they were being used in cerebral palsy with quite definite diagnostic and prognostic connotations. An example of this was the use of the term dystonia, which to Dr. Mackenzie and many Queen Square neurologists meant an abnormal state of muscle tone, and to Dr. Tizard meant the condition of dystonia musculorum deformans. In particular, there appeared to be a tendency to use single terms to define categories of cerebral palsy; thus the finding of rigidity or spasticity in a child would automatically place him in a category if some classification were used. This category in turn would have definite ætiological and prognostic connotations.

Aims of Defining Current Terminology in Cerebral Palsy—There appeared to be general agreement that the terms in common use should be clearly defined and that they should be regarded as merely descriptive. It was emphasised by Mrs. Johnson Abercrombie that the terms were "the tools of clinicians", that these tools

*Members of the Working Party: T. Bates, S. Brandt, T. T. S. Ingram, M. L. Johnson Abercrombie, R. C. Mac Keith, I. C. K. Mackenzie, P. E. Polani, W. Ritchie Russell, J. P. M. Tizard.

allowed one to describe the medical features in a given case and to communicate these clinical findings to another worker who would be able to understand the description because the terms used had been clearly defined. To classify cases of cerebral palsy it was felt, was a different problem. Categories of cerebral palsy there must be, and what criteria should be selected in establishing these categories would depend very largely on the purposes for which the classification was required. It was emphasised that no classification could be really satisfactory or universally used until the terms used to describe patients were clearly defined and understood.

Some further discussion took place on the relationship between descriptive terminology on the one hand and classification of cerebral palsy on the other. The difficulty of defining categories of cerebral palsy was generally realised. Dr. Ritchie Russell suggested that it might be possible to avoid the problem to some extent by describing patients' individual clinical features in terms of severity, using a number system. For example, hemiplegia might be described as paralysis grade 2, spasticity grade 3, athetosis grade 1, etc. The pædiatric viewpoint, however, was that a classification with broad categories of cases with definite clinical features would be of great value.

Definition of Terms.—Further discussion was predominantly concerned with the definition of various descriptive terms commonly used in cerebral palsy. In general it was felt that the current usage of terms by adult neurologists was the best guide to the way in which they should be used in the specific problem of cerebral palsy in childhood. Unfortunately we found a great deal of difficulty in obtaining agreement as to the way in which a number of descriptive terms were, or should be, used by adult neurologists, and it was apparent that this was going to be a major difficulty. It was also interesting to observe that in spite of the fact that the importance of using terms in a purely descriptive fashion had been generally agreed, all the members of the group, with the exception of Dr. Brandt and Mrs. Johnson Abercrombie and Dr. Polani, were, at one time or another, guilty of using them with a predictive, diagnostic or ætiological connotation. Dr. Ingram was particularly guilty in this respect.

Unwanted or Involuntary Movements.—Further discussion centred on the definition of the terms to describe unwanted or involuntary movements. How involuntary or unwanted movements could be distinguished from wanted movements was first discussed. Dr. Tizard postulated that there might be two forms of involuntary movement, sudden movements which occurred without reference to any particular voluntary activity and might be found when the patient was at rest and those which predominantly complicated voluntary activity but which in general showed a higher organisation in their pattern (e.g., athetosis). This was not felt to be a satisfactory guide in choosing terms to describe involuntary movements by Dr. Bates or Dr. Mackenzie.

Specific types of unwanted or involuntary movements were then discussed. The only point which appeared to be unanimously agreed was that hemiballismus occurred so rarely in cerebral palsy that its strict definition as a descriptive term was not a major problem. A considerable amount of time was spent on describing the various types of involuntary movement encountered in cerebral palsy in childhood, comparing them to those that occurred in adults. It was agreed that "chorea" should be used as a descriptive term of a rapid unwanted movement in which a considerable range of movement at one or more joints commonly occurred. Athetosis was a much slower, typically writhing, movement but it was felt that the use of this term was not confined only to the distal parts of the limb, as in the original description of athetosis by Ham-

mond. Choreo-athetosis was a term which could be used to decribe the commonly occurring involuntary movements which were intermediate in speed between chorea and athetosis and which had characteristics of both. It was suggested that the term "dystonia" was best avoided. Titubation was suggested as a suitable term to describe involuntary or unwanted movements of the head and neck. The term "tremor" was not discussed owing to shortage of time, and the group did not dicuss rigidity, spasticity, dysmetria, ataxia, epilepsy, dysarthria, aphasia, or the large number of other terms.

NOMENCLATURE AND CLASSIFICATION

GENERAL REPORT AND COMMENT BY THE SESSION RECORDER, DR. P. E. POLANI (London)

ONE WORKING party, one study group, and two general discussions of nomenclature were held at Oxford—and discussion on this topic occupied several small groups and parties. Also the clinical session brought to light several most interesting points. From all this it emerged that the greatest need was for definition of terms —not for classification. The definition of "cerebral palsy" was generally acceptable, with verbal alterations (not necessarily for the better)—and the following was agreed:

> *Definition.*—A condition characterised by a persistent impairment of control of movement and posture affecting those parts of the brain which control them and acquired in the course of the development of the brain at an early period of life.

This was modified as follows by Drs. Ingram and Polani, and the modification was acceptable:

> A non-progressive condition affecting those parts of the brain which control movements and posture, acquired in the course of early brain development.

This definition took into account various local necessities of a practical nature and also the difficulties of translation into the various languages represented at Oxford. On the classification the clinical sessions and the group discussions *seemed* to suggest a great measure of agreement, but the agreement was mostly only of appearance, not of reality. It was quite obvious that different people used the same diagnostic label with different meaning and gave it different width. Others did not appear to have thought very clearly *what* they meant by a given term, nor what its boundaries were. It was thus the meaning of the words used, even the simplest of every day neurological language, that needed definition, clarification and agreement. The term dystonia is a good example: to some, like Dr. Mackenzie, it meant an alteration of muscle tone away from normality; to others, like, I think, Dr. Ritchie Russell and Dr. Tizard, it conjured up an idea of postures fluidly and slowly running into each other, with involvement of the trunk "like in dystonia musculorum progressiva"; to others, like Dr. Ingram, it suggested the picture of slow movements that became suddenly fixed and frozen, generally in extension (like the upper limb, extended at the elbow, with fist clenched, volar flexed and pronated, the arm adducted at the shoulder, internally rotated and rigid almost behind the back)—and it sometimes suggested a "phase" in the natural history of "cerebral palsy"; to others still, like Dr. Rushworth, it suggested a particular type of hypertonus, distinguished from both spasticity and rigidity though being pillar-like, that is without a lengthening reaction.

The term athetosis is another example: to some it signified slow writhing involuntary movements; to others the notion of

"distal" was attached to the term—though they could not say what they would call a similar movement involving also the proximal segments of limbs. Others included choreic (quick) movements.

The disagreement in the meaning of terms was not only between neurologists and pædiatricians but to some extent among neurologists themselves. To add to the difficulty, it was apparent that the same term was used by the same neurologists with different meanings in different contexts. Dr. Ritchie Russell, for instance, felt that the movement pattern in "congenital double athetosis" was different from that in "athetosis" seen in adults as a result of acquired lesions—yet both were "athetosis" to him. If terms that are used for adults' disorders are applied to similar but not identical children's disorders there may be some misunderstanding.

The main task for the Little Club now is the preparation of an agreed terminology. Terms which are used in childhood neurology and are borrowed from the neurology of adults should first be defined by neurologists of adults. Such terms as athetosis, chorea, dystonia (I think this word can hardly be patched up any more), increased "tendon" reflexes, stretch reflex (if ever used by clinicians), spasticity, clasp-knife phenomenon, rigidity (cog-wheel and plastic) etc., should be strictly defined. The techniques used in eliciting physical signs and the possible alternative responses to the application of these techniques should be defined and, where possible, their inferences discussed. It should then be decided if these terms have a place in the neurology of children—and if so in what age groups—and whether the inferences that can be legitimately drawn are the same as for adults. It is probable that these terms will be descriptively meaningful only in older children, say, over five. In smaller children, in infants and newborn babies, the terms could be equivocal. For instance assessment of muscle tone requires voluntary relaxation —but can a baby or infant voluntarily relax?

It is probable that for younger infants new methods of examination and assessment, leading to a new terminology to describe signs (and symptoms) will have to be developed and that only the passing of time and careful prospective studies will tell of the correlation (if any) between infantile signs and symptoms and those of childhood (=adulthood), and hence of the "inferences" that can be legitimately drawn.

As for classification, this will be unequivocally possible only on the grounds of an agreed terminology. There is no doubt that the trend should be towards unified ætiological pictures. Some, like Dr. Ingram, have suggested neurological syndromes as a basis for classification. By this he means syndromes with a worked-out evolution, the early stages of which differ from normality and from the later (final) stages, but are predictive of these latter. This would mean that the picture of a cerebral spastic "paraplegic", at 3 months say, differs from that of a normal child and from that of a paraplegic of 3 years, yet is unequivocally and invariably "spastic cerebral paraplegia". I am sure that no case has yet been positively made for such correlations on the grounds of clinical findings alone. Thus the dystonic phase of Ingram is very much (in appearances) like the intermittent opisthotonos phase of what used to be called in a predictive way "athetoid cerebral palsy". To arrive at a classification based on neurological pictures a neurological prospective study is the only possible approach —and not one based on history, or on data collected by different people, at different times, using the apparently similar (but obviously dissimilar) criteria implied in (but not defined by) current terminology.

There are certain clinical pictures that emerge—almost clinico-ætiological correlations like:

Severe neonatal jaundice—bilateral chorea/athetosis with deafness.

Prematurity without "anoxia" (this requires definition!) — cerebral spastic "paraplegia" (diplegia with minimal involvement of upper limbs) with good intellect and freedom from seizures.

Prematurity with anoxia — diplegia double hemiplegia with intellectual (?) impairment and fits.

Postnatal infantile "cerebral palsy" — hemiplegia.

But would the diagnosis be made confidently on the physical signs alone, before the fully fledged neurological picture is revealed in the older child? The pædiatricians think this is the case in "athetoid cerebral palsy", and they say that "dystonia" and intermittent opisthotonos are clues. Is this really so? How does this dystonia differ from Ingram's dystonic phase of diplegia? I think that in fact the experienced observer can "predict" the onset of unwanted movements at an early stage of the natural history of "athetoid cerebral palsy". But I think, the pædiatrician *utilises clues other* than muscle tone: I think that, by his knowledge of "normal", he detects abnormal postures and early unwanted (possibly more truly athetotic) movements—perhaps at the subconscious clinical level. I have recently been impressed by the frequency with which I have seen athetosis in very young infants and yet I remembered the words of a colleague who had said: The normal movements of an infant are like those in athetosis! To me these movements were abnormal, and quite different from any normal infantile movements.

Finally, about classification; some workers felt that a classification should be understood, accepted and used by all sets of workers dealing with "cerebral palsy": this is foolish. There is no earthly reason why a classification—based, say, on causes, or even on neurological signs and symptoms—*should* be a good basis for a treatment classification: it may be, it need not be. If for treatment then why not for education? A classification of use in planning treatment *may* coincide with a clinical neurological classification—this could be so if treatment were based on neurophysiological principles and if signs and symptoms could invariably describe upsets of neurophysiological mechanisms. All told, classification, on the clinical plane, should be based on clinical findings from which meaningful inferences to pathology and physiological mechanisms could be made. In a second stage, correlation with causes may become established and a causative classification may be adopted. At this stage a treatment classification may easily cut across ætiological boundaries—and so might classifications to meet other practical needs (education, training, etc)

Recommendations:

(1) The Little Club should have discussions on terminology at two levels:
   (a) between neurologists;
   (b) between them and pædiatricians with multiple practical demonstrations on *few* cases.
(2) A philologist should be present.
(3) We should aim at definitions of terminology and of techniques.
(4) Having got terminology and techniques right (i.e., exactly and unequivocally described), experienced, unbiased workers should examine children with "cerebral palsy",
   (a) at schools;
   (b) in institutions for mentally defectives;
   (c) in children's hospitals or in their O.P. departments (=babies!).

One (or more) examiner(s) should carefully examine these patients and see if the physical signs described in the "terminology" did apply. If so, all well and good; if *not* then descriptions, not names, should be used. Perhaps new groups of findings would be demonstrated, similar (perhaps) to those defined but not identical with them and requiring a different name. My guess is that this would happen often. A practical point: every patient should be examined care-

fully—and repeatedly, and a secretary should take notes dictated during the examination. If more than one observer could be found, all discrepancies between observers should be sorted out by joint examination of the same patient. A hundred or more patients should be examined, with ample time for thought and discussion—and perhaps something new and meaningful might emerge—histories should not be available until the end of the study.

Following the examination of the hundred patients, classification of cases according to signs should be possible, and later they could be correlated with pathological findings, neurophysiological mechanisms, causes, etc.

What I am saying, in fact, is that infant neurology is a new clinical science which requires new techniques of examination and often new descriptive terms —terms which cannot always be found in the armamentarium of the neurologists of adults.

PAUL E. POLANI.

GENERAL DISCUSSION of the Little Club Memo

DR. RITCHIE RUSSELL (*Oxford*)—It is a great mistake in a difficult and changing situation of this kind to attempt to introduce any uniformity. It is much better that the special units who are studying this subject should establish their own system of classification, making it quite clear what they are talking about; then if their's is the best method other people will make use of it.

With regard to the term "double hemiplegia"; in adult neurology I think this tends to be used when the lesion has been caused by two separate episodes. I'm not very happy about using it for a condition which was caused at birth by a single thrombosis, or whatever it was, but if it came on in two episodes then one could use it—just as one could use the term "double hemianopia" for a cortical blindness, if it occurred on two separate occasions.

DR. WILLIAM DUNHAM (*London*)—I have heard several references this morning to "variable rigidity". This syndrome has been referred to frequently in this set of meetings, as cerebral palsy of the mixed type. We can talk about mixed ætiology and we can talk about a combination of physical signs, but I am not sure that we can talk about mixed diagnosis. We have here a separate syndrome to describe, and it is useful, for what is now called a mixed type, to have a separate name, which is just a convenient way of referring to a list of signs and symptoms.

In the variable rigidity syndrome, originally described by Eirene Collis, the tonic reflex positions are highly characteristic. The involuntary head movements and dystonia of the limbs and trunk are striking signs. But it is worth noting that in this syndrome not only are tonic neck reflex arm position elicited by rotation of

the head, either active or passive, but the head position is also influenced by the position of the arms. If, when a child with this dystonic cerebral palsy is being handled, both his arms are moved simultaneously and symmetrically, the head tends to remain in the midline, a fact which is of great value to those undertaking the general management and training of the child. For this syndrome you have the qualification which Mackenzie mentioned; it is a diagnosis which has practical value for the handling of the child.

The Chairman, DR. E. A. CARMICHAEL (*London*)—When I started neurology I came under the influence of a teacher, who told me: "Never use any words that are from another language; describe exactly what you do, exactly what you see, in simple language and there will never be any doubt what you mean." That is what we all must do. The object of this Memorandum was to try and get some agreement about terminology which would be used in describing cases of cerebral palsy. It is very important that any terminology that we do use should be simple, clear, and preferably good Anglo-Saxon, which can be translated into almost any other language and leave the meaning quite clear. The speakers have helped to clear the air. Diagnostic labels are a telegraphic form of communication; perhaps we have not yet reached a stage when we can use it. We should just state clearly and in simple terms what we do and what we observe.

DR. R. C. MAC KEITH (*London*)—There are various ways in which the Memorandum might be useful. One was to make clear to others our meaning of the terms we were using in this document. The second way was to put forward the method we actually use in our minds in differentiating and so classifying any individual case. We look at a group and we remove the spastic cases and then we remove the ones with obvious unwanted movements, and then we recognise what we have called the dystonics and then the ataxias. These were points to put down because when anyone starts in this field he experiences difficulty in finding any way set out for approaching the differentiation or classification of his cases. The next point which is very near to our heart was that we owe so much to the neurologists that we wanted to return to cooperation and partnership with them; that was an underlying motive in working out the memorandum.

DR. P. E. POLANI (*London*)—Professor Illingworth says that the classification of the American Academy is almost universally accepted. I know of nobody in this country who uses it and nobody who really pays more than lip service to it. Professor Illingworth could have said that everybody uses the terms rigidity, athetosis, spasticity and so on, which are the basis of the classification. But those are names, and if people use them with different meanings they are not following the classification of the American Academy. I do not see how anybody can follow that classification satisfactorily because it does not explain clearly enough the meanings they give to the terms they use. To say that the characteristic of spasticity is the presence of a pathological stretch reflex, without describing how the tests for this are done, appears to me confusing. For such reasons I do not think that either this or any other classification is universally followed. In his new book, Professor Illingworth says he follows the American classification, but though he uses the same terms he does not always follow it.

MR. G. A. POLLOCK, F.R.C.S.E. (*Edinburgh*)—I should like to support Professor Illingworth's defence of the American classification which I follow also. I don't for one moment claim that it is perfect, but, as a clinical classification, it enables one. from the clinical point of view, to file cases for one's own use and the use

of those trained in the same school. In our present state of comparative ignorance regarding the ætiology of C.P., perhaps we are paying too much attention to its classification and terminology, though a universally accepted terminology and classification is needed. It would allow the other fellow to know what is being done and might also act as a guide to treatment. My successes during the last ten years of treating cerebral palsy have been unimpressive, and I think if we all told the truth we'd be forced to admit the same. I suggest that we do not rush into a new terminology and classification at the moment, in the hope that within the next year or so research may put us in possession of more exact knowledge of the ætiology and pathology of this condition. Based on such information, a new classification and new terminology would receive instant acceptance. Until we have factual knowledge of the causes of cerebral palsy, and of the regular changes which take place in the brain structure, I suggest we continue to use a clinical classification.

BOOKS—New and Not So New

MENTAL DEFICIENCY: THE CHANGING OUTLOOK. Ann M. Clarke and A. D. B. Clarke. London: Methuen, 1958, *pp.* 513.

This is a really valuable book. It covers perhaps more of the theory of mental deficiency than the other excellent book on the subject by Hilliard and Kirman. Both books must find their place on the shelf of anyone concerned with mentally defective children, and certainly of pædiatricians.

The two authors have been assisted by J. Tizard, N. O'Connor, Elspeth Stephen, H. C. Gunzburg, and Margaret Penwill. The book contains chapters on the prevalence of mental defect; the classification of mental deficiency; the measurement of intelligence; genetic and environmental factors; the ætiology of mental deficiency; physical characteristics; learning difficulties and processes; brain damage; cerebral palsy; psychological assessment; educational problems; the trainability and rehabilitation of imbeciles and feeble minded persons; psychotherapy; speech disorders and speech therapy; and longitudinal and follow-up studies. There are 750 references, which include the full titles of the papers. Many of these references are not normally accessible and are therefore all the more useful to know about. At the end of the sections and chapters there is a good summary.

All the authors have given a thoroughly critical review of the work of others. They have not merely quoted their work, but have weighed up and assessed its value.

One cannot pick out any one chapter as being better than any other; but the sections on environmental and genetic factors are particularly useful. The well-known work of the Clarkes on rehabilitation is included here and amplified. Its importance cannot be over-stressed. The chapter on cerebral palsy is a brief review of the subject. It is accurate, so far as it goes, a possible exception being the statement that the pathological lesion in ataxia is in the cerebellum.

The authors are to be congratulated on this excellent book, which will surely be a classic in the field.

R. S. ILLINGWORTH

ABSTRACTS

IN COLLABORATION WITH "*Abstracts of World Medicine*," PUBLISHED BY THE BRITISH MEDICAL ASSOCIATION

Prematurity and "Cerebral Palsy"

P. E. POLANI. *British Medical Journal*. Dec. 20, 1958, **ii**, 1497-1499. 42 refs.

The author presents a review of the literature dealing with the association between prematurity and cerebral palsy, which is defined as "a persisting qualitative motor disorder due to non-progressive damage of the encephalon occurring before the growth of the central nervous system is complete". The figures cited all support the view that the incidence of prematurity is much higher in infants with cerebral palsy than in a comparable group of controls. Cerebral palsy, like prematurity, is an aggregate of clinical syndromes of varied origin. The factors causing prematurity act on the fœtus at different stages and with varying intensity, while birth weight affects the incidence of cerebral spastic paraplegia. It has been shown that in premature infants weighing less than 4 lb. at birth the chance of developing cerebral paraplegia or diplegia is about 1 in 20.

Another study has shown a high degree of correlation between prematurity and dystonic/choreo-athetoid cerebral palsy following severe neonatal jaundice not due to blood group incompatibility, premature babies having a greater tendency to a rise in the serum-bilirubin level, presumably because of the inability of the premature liver to conjugate the liposoluble bilirubin with glucuronic acid to form water-soluble pigments I and II, with the result that the liposoluble pigment crosses the blood-brain barrier and produces kernicterus. This process may be increased by the administration of excess vitamin-K analogues or sulphafurazole, an agent which is known to favour kernicteric damage, possibly by lowering the threshold of the blood-brain barrier. Other mechanisms postulated include "anoxia" and mechanical trauma leading to brain damage, but this is not acceptable in view of the striking symmetry of neurological signs. It is considered that in only a small proportion of cases are obstetrical factors responsible for anoxia leading to cerebral palsy. If a developmental mechanism is at work then the cause of cerebral palsy may well be the same as the cause of the prematurity. Lilienfeld and Parkhurst postulated a "continum of reproductive wastage", with cerebral palsy as one link, and other elements, such as mental defect, epilepsy, behaviour disorder, and lowered intelligence, all representing the results of a harmful association between prematurity and brain damage

Recent experiments suggest the possibility of a hyperoxic mechanism, such as can happen in retrolental fibroplasia. The entirely hypothetical suggestion that "premature birth may at times cause post-natal interference in development and maturation of critically maturing neurones" can be put to the test by carrying out experiments with high oxygen pressures in immature animals and observing the results. Such an hypothesis directs attention to the possibilities of prevention.

M. R. Medhurst

Electroencephalographic and Clinical Study of Anoxic Convulsions in Children. Their Location within the Group of Infantile Convulsions and Their Differenciation from Epilepsy

H. GASTAUT and Y. GASTAUT. *Electroencephalography and Clinical Neurophysiology*. Nov., 1958, **10**, 607-620. 3 figs., 23 refs.

Writing from the University of Marseilles, the authors express the view that convulsions in infancy are often due to episodes of cerebral anoxia and not, as is often assumed, to an epileptic process. In a study of 176 infants and children aged a few months to 5 years, in whom adequate clinical details concerning the attacks were available, the electroencephalogram (EEG), electrocardiogram (ECG), and respiratory amplitude and rate were recorded simultaneously

during the waking state, during natural sleep, and also during photic stimulation and hyperventilation (where possible). In each case the ocular globes were compressed strongly for a period not exceeding 10 seconds, the procedure being sometimes repeated 2 or 3 times. This manœuvre produced anoxic convulsions in 24 children of a mean age of 28 months—that is, in 13·5% of the cases. In these 24 children the EEG, ECG and respirogram were normal before ocular compression, but in each case marked changes in cardiac and/or respiratory rhythm were observed during compression, these changes being accompanied by paroxysmal slow activity in the EEG.

Neurologists and pædiatricians are often unaware of the possible anoxic mechanism concerned in childhood convulsions. Many so-called essential, hyperthermic, and even afebrile convulsions in childhood are due to this mechanism, and breath-holding seizures in particular are certainly due to this cause. Polygraphic recordings of the type described may identify the nature of these attacks. The long-term prognosis is very much better in such cases than in those in which the attacks are due to an epileptic mechanism.

John N. Walton

Education of Cerebral Palsied Children. The Role of Meprobamate: a Preliminary Evaluation

B. E. KATZ. *Journal of Pediatrics*. Oct., 1958, **53**, 467-475. 11 refs.

Children with cerebral palsy have two major groups of symptoms which handicap their education: (1) purposeless body movement with spasticity; and (2) emotional difficulties leading to abnormal behaviour. Both these symptom complexes must be treated to allow full intellectual development. Meprobamate, a muscle relaxant capable of relieving anxiety and emotional tension, has been shown to lessen behaviour disturbances, and the present study was undertaken to assess the influence of the drug on learning ability.

Of 19 patients attending a school for children with cerebral palsy, 10 were selected for treatment and their progress compared with that of the remaining 9. The ages of the 10 treated patients ranged from 5 to 24 years and their I.Qs from normal to below 60 on the Terman-Binet scale. The degree of physical handicap varied from mild to severe, and behaviour was mildly or moderately disturbed in 5 cases and normal in 5. One patient could not read and the other 5 had varying reading abilities. All were physically healthy, and all had been under observation for at least 2 years. [Corresponding details in respect of the 9 untreated patients are not given.] Meprobamate was given daily in a dose of 200 mg., increasing by 200 mg. fortnightly until a satisfactory response was obtained, treatment being discontinued during the holiday and at weekends unless the parents asked for it to be continued, as they did in 6 cases.

A weekly assessment was made of: (1) degree of muscle relaxation; (2) magnitude of spastic or involuntary movement; (3) ease of handling; (4) outlook and attitude; (5) endurance; (6) attention span; and (7) ability to learn. Evaluation of the first three was performed by the medical officer and physiotherapist and followed closely the standards defined by Gillette (*Int. Rec. Med.*, 1956, **169**, 453; *Abstr. Wld. Med.*, 1956, **20**, 474) and of other authorities where applicable. The last three factors were evaluated by two teachers, and the patients' outlook and attitude by all four observers. Details of the response of each subject to various doses of the drug are tabulated.

The 9 patients not given meprobamate continued at the previous retarded rate, while those treated became more manageable both physically and emotionally during the 80 weeks of the study. They were less fatigued and more alert, while in 9 out of the 10 accelerated reading progress and ability to learn were observed, probably attributable to reduced spasm and better concentration, with increased attention span. The dosage of the drug had to be adjusted carefully to each child's specific requirement, the maximum level varying from 400 to 1,600 mg. daily.

It is not claimed that the results prove that meprobamate has a direct effect on attention span or learning ability, but they do show that the response to instruction of the children studied improved during the period of the study, and the author considers that meprobamate was responsible for this unexpected level of progress.

M. R. Medhurst

Learning Disabilities Associated with Lesser Brain Damage

H. E. THELANDER, J. K. PHELPS, and E. W. KIRK. *Journal of Pediatrics*. Oct., 1958, **53**, 405-409. 4 refs.

Children with cerebral palsy and other evidence of brain damage have been studied over a period of 5 years at a special unit of the Children's Hospital of San Francisco. This consists of an admission clinic, a nursery school, a special evaluation service, and a follow-up clinic. The children are observed by both teachers and psychologists. In addition to the usual standardized intelligence tests and developmental scales, special tests are used to assess perception, visual-motor coordination, auditory memory span, and qualitative approach to tasks. These tests often have to be adapted to the child and are therefore not standardized, but an attempt is being made to correlate the results of various tests with known brain damage and particular patterns of behaviour.

Seven cases are quoted to illustrate how minimal motor disturbance may be accompanied by specific perceptual defects which lead to difficulty in learning. Ordinary neurological examination, electroencephalography, and air encephalography are too insensitive to detect such abnormalities and need to be supplemented by specially devised tests carried out by skilled personnel. The more subtle defects which lead to learning difficulties are those of: (1) expressive speech; (2) understanding of oral commands; (3) writing; (4) perception of hearing; (5) memory; (6) control of hyperactivity; and (7) emotional control.

Children with cerebral damage causing aberrations of this type, but with minimal motor involvement, merge with those who have no demonstrable motor handicap and thus tend to be excluded from the category of cerebral palsy. If their deficits are not understood and treated by an intensive rehabilitation programme serious emotional maladjustment may result.

M. R. Medhurst

Long-range Effects of Electropallidoansotomy in Extrapyramidal and Convulsive Disorders

E. A. SPIEGEL, H. T. WYCIS, and W. H. BAIRD III. *Neurology*. Oct., 1958, **8**, 734-740. 1 fig., 24 refs.

The authors report the results of a long-term follow-up of patients treated for various neurological conditions by stereotaxic operations on the basal ganglia. In the majority of cases areas of the pallidum and ansa lenticularis were electrically coagulated, while in some epileptic patients additional coagulation was produced in the amygdaloid nucleus. The operations were carried out under local anaesthesia whenever possible so that the effects of both stimulation and coagulation on the symptoms and signs of the disorder could be assessed.

Of 50 patients aged 19 to 79 years suffering from postencephalitic Parkinsonism or paralysis agitans of 3 to 34 years' duration, subjected to 71 operations, bilateral in 9 cases, and followed up for 1-6 years, the tremor which had been unresponsive to medication was abolished or greatly reduced in 22 (44·9%) and was moderately improved in 16 (32·6%). In 6 cases, however, the tremor recurred a few days to 20 months after operation; these were patients in whom the lesion had been made anteriorly in the pallidum, either at the level of the anterior commissure or 3 mm. posterior to it, and when in 5 of these the lesion was enlarged posteriorly up to 9 mm. behind the anterior commissure the tremor was abolished in 4 and reduced in one. Rigidity, which was a marked feature in 36 of the patients, was greatly reduced in 26 (72·2%). There were 2 deaths. Permanent hemiplegia occurred in 2 cases (4%) and transient paresis in 6 (12%). These complications were commonest among the older patients. Complete rehabilitation enabling the patient to resume previous employment was obtained in 9 cases (18%) and partial rehabilitation in 15 (30%).

Further groups of patients with other extrapyramidal conditions included 4 cases of Huntingdon's chorea, in which involuntary movements were abolished in 1 case and reduced in 2, but 2 patients with other types of chorea were unchanged. In athetosis it is advisable to test the probable effects of coagulation by giving a preliminary injection of procaine, because there is a possibility of making the condition worse by pallidotomy. In one case of post-hemiplegic athetosis the movements were abolished; in 2 cases of double athetosis improvement was obtained on the worse side by a unilateral operation; but in 2 cases of cerebral palsy with athetosis only slight improvement was

obtained, while of 2 cases of torsion dystonia, one was greatly improved and the other slightly improved. As regards the treatment of spastic lesions, in 2 cases of hemiplegia no reduction in spasticity was noted, but in a third case in which a sudden hemiplegia had followed ectopic pregnancy the spasticity was greatly reduced following operation, and tonic innervation, which had been a particularly disabling feature, was abolished.

Lastly, in 12 patients with epilepsy, 5 treated by pallidotomy and 7 by pallidotomy and amygdalotomy, and followed up for periods of 6 months to 2 years, the attacks were controlled or markedly reduced in 7 (58·3%), but in the remainder the attacks were unchanged or returned after temporary improvement; in all but 2 of these cases, however, the operation was unilateral only. "Salaam" seizures were most favourably affected, being abolished in 5 out of 8 cases and greatly reduced in number in 1.

Brodie Hughes

Cerebral Palsy. A Clinical Study of 370 cases

MARIT SKATVEDT. *Acta Pædiatrica.* Nov., 1958, **47**, 706-713 (Summary of Suppl. 111.)

The histories and clinical findings in 370 children with C.P. have been analysed to illustrate the ætiology. The ratio of males to females was 6 to 4, and there was a significant predominance of first-born children (52·7%). No evidence was obtained of maternal illnesses or other external factors that might have damaged the fœtus during pregnancy. Abnormalities in labour were noted in 44% of the cases, protracted labour and breech presentations being the main ones. In 47% of the children there was a suggestion of perinatal asphyxia and/or symptoms of cerebral injury. In 26 children (7%) there was evidence of kernicterus in the neonatal period.

The incidence of prematurity (birth-weight under 2,500 g.) was 28%. Of the children with bilateral spasticity, 49% were prematures, a relatively large percentage having a very low birth-weight. On the other hand, there was also a significantly high proportion of large infants weighing over 4000 g. at birth. The percentage of normal birth-weights was therefore considerably lower than in the general population (55% against 80-85%).

There were 25 pairs of twins, in 2 of which both infants had C.P.; in 6 cases the other twin was born dead, and in 15 it was apparently normal.

In 18 cases (5%) there was a possible postnatal cause for the C.P.—either cerebral infections or other cerebral injury. There was no evidence of the ætiological importance of hereditary genetic factors or external fœtal injuries. But there was some evidence pointing to a dominant importance of perinatal brain injuries in the causation. In a comparatively small group these cerebral injuries are due to kernicterus. This condition is not rare in prematures without any blood-group incompatibility. It is suggested that mild cases of kernicterus may occur and pass unnoticed in the neonatal period, leading to C.P. This view is supported by the frequency of prematurity and of clinical symptoms of asphyxia/cerebral hæmorrhage in this study. More significant is the high proportion of pathological deliveries and of infants weighing over 4000 g. at birth. It must be presumed that the same cerebral injuries that cause perinatal deaths can produce C.P. when they are of sublethal degree. The prevention of C.P. must aim at reducing these injuries.

The cases are classified into three main types—uncomplicated spasticity (57%), with the subgroups of spastic hemiparesis (24%) and spastic diplegia (33%); uncomplicated athetosis (14%); and uncomplicated ataxia (6·5%). The remaining 22% predominantly presented a mixture of pyramidal and extrapyramidal symptoms.

Pneumoencephalography, done in 105 patients, suggested that there is a correlation between the degree of PEG changes and of motor and mental defects. The PEG in C.P. strikingly resembles that in epilepsy without neurological symptoms.

Organic epilepsy, C.P., and mental impairment due to cerebral lesions should be considered as one pathological entity with a common ætiology and anatomical basis.

Ed. C.P. Bulletin

The annual subscription to this Bulletin is £1 ($3.25; 14fr.; 12fl.; 20kr.); *including inland and overseas postage, payable to the Secretary, National Spastics Society,* 28 *Fitzroy Square, London, W.*1.

Correspondence should be addressed to the Editorial Board at 28 *Fitzroy Square, London, W.*1, *or to the Editor at* 29 *Hadlow Road, Tonbridge, Kent.*

Back numbers can be supplied at 5/- *per copy, post free.*